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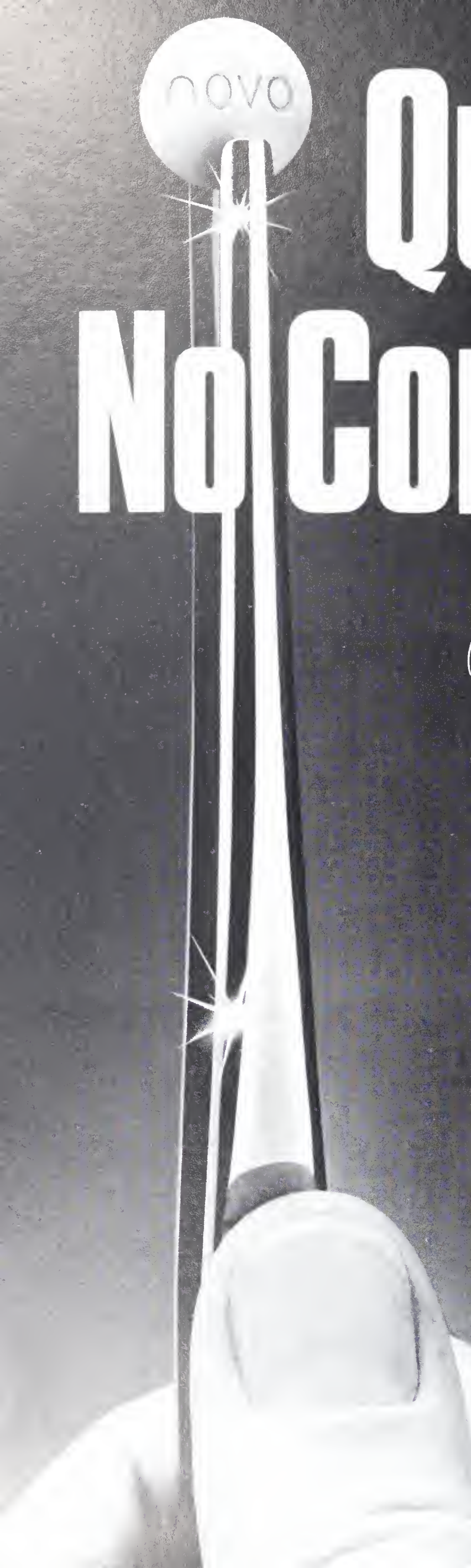

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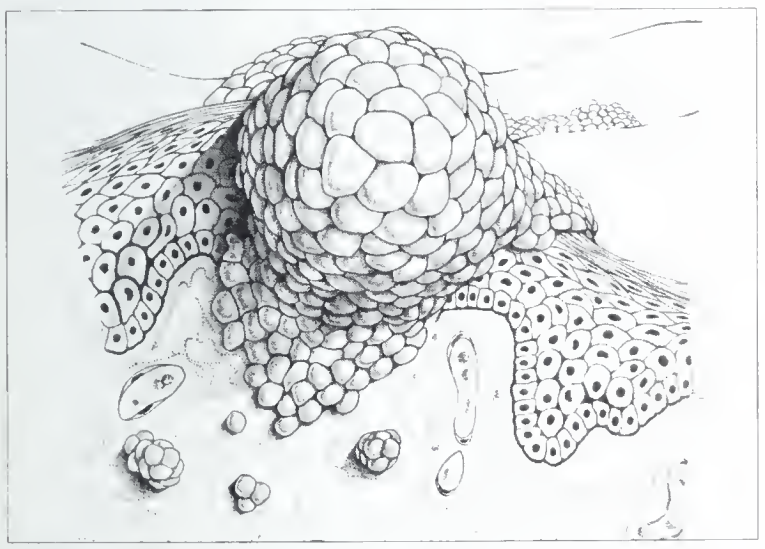

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Editorial

Founded in 1923, the University of Toronto Medical Journal is celebrating its 69th year of publication. As editors, we are fortunate to have a number talented individuals on our editorial board and as contributing authors. This year's editorial staff was selected from a number of well-qualified candidates. We feel our board reflects the wide base of knowledge and interests characteristic of the University of Toronto's undergraduate medical community. Gino Maulucci, as art director, has been instrumental in the design of the UTMJ which features the work of artists from the Department of Surgery's Division of Biomedical Communications (formerly the Department of Art as Applied to Medicine). Illustrations from students in the BScBMC 9T3 program may be found on the cover of this issue and accompanying each article.

In this issue, Neal Riekenbrauck and colleagues discuss a spindle cell variant of malignant melanoma and Willoughby et al provide an interesting review and case report on colorectal carcinoma. In the feature article, Ariel Waitzman and Jeffrey Posnick describe the surgical correction of orbital hypertelorism. The article demonstrates the striking results which can be achieved through modern craniofacial reconstructive surgery and illustrates the importance of the team approach required for management of patients with this physical finding.

It is our pleasure to publish the winning essay of the third annual Biomedical Ethics Essay Competition. In their award-winning paper, Gil Aronson and Dan Farine consider the medical, legal and ethical aspects of maternal-fetal conflicts. Diagnostic advances and developments in fetal therapy have served to enhance the status of the fetus - differentiating the fetus from its mother. The authors discuss current efforts to deal with maternal-fetal conflicts and call for a framework which may be used to resolve conflicts in the future.

Also in this issue, Steven Friedman provides clinical and pathological perpectives on Oral Lichen Planus, and Michele Carr and Amis Freiberg discuss the diagnosis and management of carpometacarpal joint osteoarthritis. Quick Diagnosis, News & Views and Book Reviews round out the issue.

The UTMJ serves as a forum for students to communicate research and clinical experiences. We encourage students to write articles in both basic and clinical sciences. As in previous years, the UTMJ will award several prizes to the articles published in this Volume:

The best article as selected by the editorial staff will recieve the **University of Toronto Prize for Literary Excellence,** consisting of $100. Second and third place articles will receive $50 each.

The best article in the field of orthopaedics as selected by the Department of Orthopaedics will receive the **R.I. Harris Award.**

Dr. Richard Gladstone will award the best article in the neurosciences the **Gladstone Award,** consisting of a certificate and $100.

The winners of the awards will be announced in the third issue. We hope these will serve as a further incentive for students and members of the University of Toronto medical community to publish their work.

In order to improve the quality of the UTMJ we would appreciate feedback from our readers. We are planning to include a 'Letters to the editors' section in upcoming issues. Please send us your comments, submissions or concerns regarding the UTMJ and its contents.

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Maternal-Fetal Conflicts: Medical, Legal and Ethical Aspects

Gil Aronson 9T2, Dan Farine, M.D., F.R.C.S.(C)

Recent technological advances provide clinicians with the ability to diagnose many fetal disorders in utero.[1] Diagnostic advances have enabled the development of medical and surgical therapies for fetal disease. The difficulty with these therapies is that they differentiate the fetus from its mother, setting up potential maternal-fetal conflicts. Presently, these conflicts are being solved by some physicians who use medical and legal force against the mother's will. Unless a framework for resolving conflicts is developed now, emerging treatments will lead to further situations in which the mother's right to refuse treatment is overridden by her physician and the courts. Hence, the different aspects of maternal-fetal conflicts need to be examined in order to prevent future disputes.

Medical aspects

Prior to 1960, prenatal diagnosis depended on physical examination through the maternal abdominal wall. It was only during the 1960's that we began to learn more about the fetus, as amniocentesis became available to women of advanced maternal age for the diagnosis of genetic and biochemical abnormalities. With the advent of real-time ultrasound, the fetus could be further scrutinized for deformities. Soon, these prenatal diagnostic techniques became part of search-and-destroy missions, as abnormal fetuses were aborted to avoid birth defects. More recently, fetal therapy has become a viable option to termination in the presence of prenatal disease. As a consequence, the status of the fetus in the medical community has increased.

Fetal therapy can be administered both directly and indirectly to the fetus. Indirect therapy requires venipuncture of the pregnant woman and may be used for either prophylaxis or treatment of disease. The first indirect therapy to be used involved the administration of Rh immune globulin (RhIg) to Rh-negative women in order to prevent Rh isoimmunization in subsequent pregnancies.[2] In addition, respiratory distress syndrome of the neonate has been prevented through maternal injection of glucocorticoids, which promote fetal pulmonary maturity.[3] The fetus may also be medically treated through the mother if there is sufficient transplacental passage of the medication. For instance, methylmalonic acidemia has been treated maternally with large doses of vitamin B12,[4] while multiple carboxylase deficiency has been managed with maternal biotin during the last half of pregnancy.[5,6] Also, in cases of suspected congenital adrenal hyperplasia, the fetal adrenals have been successfully suppressed by dexamethasone therapy to the mother.[7,8]

In 1963, Liley performed the first direct fetal procedure by transfusing a fetus suffering from severe erythroblastosis fetalis.[9] Further direct techniques for treating fetal diseases have since stemmed from fetoscopy and ultrasound-guided needle insertion. Currently, these invasive techniques are being used to visualize the fetus, sample its tissues, usually blood, and treat any diseases. For instance, fetal transfusion is now done by the percutaneous injection of blood, through the uterine wall, into the umbilical vein.[10] Severe hydrops caused by this disease can be treated in the same injection with digitalis and diuretics.[11]

Some of the congenital anatomic lesions, that interfere with normal development, have also been treated percutaneously in an attempt to prevent complications. One of these lesions, urethral obstruction may lead to hydronephrosis, cystic renal dysplasia, oligohydramnios, pulmonary hypoplasia and abdominal wall muscle deficiency.[12] If the urine is drained from the bladder using a chronic vesico-amniotic shunt, these complications can be prevented. However, this approach has been successful in only 40% of 87 cases.[13] Therefore, controlled trials are needed to determine if there is an approach with better efficacy and survival. The treatment of obstructive hydrocephalus, due to aqueductal stenosis, has been even more disappointing due to significant morbidity and procedure-related fetal mortality.[14] For these reasons, there is presently a moratorium on in utero shunting for hydrocephalus until a more effective approach is found and patient criteria are better delineated.[15]

Open fetal surgery is both the latest and the most invasive technique available to reach the fetus. It requires a hysterotomy or classical cesarean incision so that the fetus can be accessed. This procedure puts the mother at risk, dooms her to have cesarean sections for her remaining pregnancies and increases her risk of uterine rupture. Diaphragmatic hernia, a grave condition causing pulmonary hypoplasia and death, is the most recent fetal disorder to be treated by open surgery. The surgery involves repair of the diaphragmatic defect and enlargement of the abdominal cavity by abdominoplasty to accommodate abdominal organs.[16] Future open surgical corrections may also be possible for spina bifida[17] and cleft lip and palate.[18]

Promising future fetal therapies, that are already being attempted, include hematopoietic stem cell transplantation and gene therapy.[19] Stem cell transplantation may be an effective prenatal treatment for hematologic disorders due to the normal delivery of stem cells from the fetal liver at twenty weeks and fetal immunotolerance to allogenic grafts.[19] Somatic cell gene therapy is currently being investigated for the replacement or correction of defective mutant genes. If given prenatally, this gene therapy may prevent the metabolic consequences of some of these defective genes.[20] Research is also being done on germ-line gene therapy, which involves the microinjection of a new germ line into the fetus' cells. However, this therapy has many ethical problems since it involves the permanent modification of the human genome. Technically, microinjection has high failure rates and may lead to the production of pathological conditions.[21] As techniques, such as embryoscopy, are developed to view the fetus at an earlier stage, the barriers to cellular and genetic therapy in the fetus will be eliminated.[22]

Legal aspects

As legal interventions sought by physicians and hospitals have increased over the last decade, it has become more evident that the courts are poorly equipped to handle these cases. According to Kolder et al,[23] judgements are often requested on extremely short notice, leaving insufficient time for both sides to find adequate and prepared counsel. Similarly, judges do not have enough time to become familiar with the medical and legal problems involved and to reflect on any past precedents and future impacts.[24] Therefore, decisions are rushed, impulsive and lack well-reasoned conclusions. As many of these hearings occur in the hospital, judges tend to agree with the physician's medical advice.[24] In the end, the pregnant woman is left without the opportunity for appeal.

Most of the cases of obstetrical interventions that have been authorized by courts of law have been based on the U.S. Supreme Court's decision on abortion, *Roe vs. Wade*.[25,26,27] Ironically, this is the same ruling that asserted the right of a woman to make her own decisions concerning pregnancy. According to the *Roe* decision,[25] the state has a compelling interest in protecting the fetus, once it has reached the point of viability. Thus, the state is allowed to prohibit abortion at this time. Courts have interpreted this 'compelling interest' as the ability to force a pregnant woman to undergo intervention in order to protect her viable fetus.[27] However, these courts have ignored the fact that the Supreme Court limited the state's 'compelling interest' in *Roe* by emphasizing that the woman's life and health come first even after the fetus is viable.[25] In addition, one must realize that simply because the state can prohibit fetal destruction, does not imply that it can force major interventions to preserve the fetus.

In more recent abortion decisions, the Supreme Court has reiterated that the woman's health must always prevail over the fetus' life and health when they conflict.[28,29] The Court invalidated state abortion statutes that required a woman having a late abortion to bear an increased risk to her health in order to benefit her viable fetus. In coming to its decision, the Court found that the trade-off between a pregnant woman's health and fetal survival was unconstitutional.[29] Since all obstetrical interventions involve this trade-off, lower courts cannot force pregnant women to undergo unwanted medical treatment, even if this treatment may benefit the fetus.

In other cases, child neglect laws have been interpreted to apply to fetuses too.[30,31] According to this interpretation, the pregnant woman could not deny her fetus the right to proper prenatal care. While these laws are based on parents' legal duties to their children, pregnant women do not have any legally enforceable duties to their fetuses.[26] Hence, these women cannot be charged with fetal neglect. It is also unclear whether fetal and child neglect are similar since treating a child against the parents' wishes is not difficult, while treating a fetus requires invasion of the mother's bodily integrity. In fact, two higher courts have stated that a court cannot assert control over a fetus, as if it was a child, even if there was neglect by the parents.[32,33]

Most of the cases of court-ordered obstetrical interventions have been lower court decisions and have not set precedent.[34] Of the three court orders that were appealed to higher state courts, one was overruled and two were upheld. The earliest decision to uphold a court-ordered obstetrical intervention, *Raleigh Fitkin-Paul Morgan Memorial Hospital vs. Anderson*,[35] was decided by the New Jersey Supreme Court in 1964 before *Roe vs. Wade* established the rights to self-determination and bodily integrity. In the other decision that upheld an ordered intervention, *Jefferson vs. Griffin Spalding County Hospital Authority*,[30] the Georgia Supreme Court denied the mother's constitutional right to refuse treatment. In coming to its decision, the Court gave little explanation or policy discussion, except to state that they were unaware of any previous child neglect judgements in which the child was unborn. In the most recent decision on court-ordered interventions, *In re A.C.*,[36] the D.C. Court of Appeals, sitting in full, overruled its previous decision that forced a pregnant woman to have a cesarean section. In its new ruling, the Court set strong legal precedent by stating that "in virtually all cases the question of what is to be done is to be decided by the patient, the pregnant woman, on behalf of herself and the fetus" since "a fetus cannot have rights in this respect superior to those of a person who has already been born."[36] The Court also added that a case would have to be quite extraordinary for a court to justify overriding the patient's decision.

Ethical aspects

The physician-patient relationship in medicine is built on the ethical principles of beneficence and autonomy. According to the principle of beneficence, the physician's actions must always help the patient and be in the patient's best interests. However, beneficence does not necessarily imply that the physician can override the patient's right to individual autonomy. Autonomy gives the patient the privilege of accepting or rejecting medical advice based on personal, religious and societal convictions without interference from others.[37] If the patient is a pregnant woman, she will make her decisions based on her convictions and her responsibilities to her fetus.

Since decisions that result in health risks are only properly made by the patient that bears those risks, the patient's informed consent is required before any type of intervention takes place. Informed consent implies that the physician must respect the wishes of adults, who are mentally competent, in situations of medical decision making.[38] In obstetrics, these wishes may be quite frustrating if the pregnant woman is noncompliant, since her decisions also affect the fetus.

However, attempts to legally force compliance raise many ethical problems since they attempt to alter the informed consent doctrine by circumventing the mother's decisions and allowing the court to decide the mother's and fetus' fate.

In situations where maternal-fetal conflicts arise, the woman's right to autonomy must be weighed against the potential benefit to the life and health of the fetus. In certain cases, such as taking a prenatal vitamin, the benefit for the fetus far outweighs the risk for the mother. This imbalance leaves the mother with a moral duty to act on behalf of the fetus. However, the physician, being responsible for the health of both mother and fetus, is obligated to act in both of their best interests. This dual responsibility implies that the physician cannot balance the interests of the fetus against the interests of the mother. Therefore, the physician must respect the woman's wishes and not override her right to reject personal risk, especially since there are still doubts as to the accuracy of some of the diagnostic procedures.[39]

The development of fetal therapy has enhanced the moral status of the fetus. It has allowed the fetus to attain the status of patient, without quite earning the status of personhood. However, the pregnant woman, by deciding not to have an abortion, has allowed the fetus to obtain that status of personhood with limited rights. Hence, her choice to continue with her pregnancy leaves her with a moral duty to protect and not harm the fetus that she is carrying in her womb. If she chooses not to comply with her physician's advice, she is inflicting harm on her fetus and is morally wrong. However, her moral duty is not absolute and does not imply that a pregnant woman, by deciding not to abort, is subordinating her right to autonomy to the fetus' right to life.

Social aspects

In several of the cases of court-ordered obstetrical interventions, the pregnant women fled the hospital and gave birth elsewhere.[26] If pregnant women are further forced by the law to comply, they will either withhold crucial information to prevent self-incrimination or avoid prenatal care entirely.[40] Hence, the majority of fetuses will suffer for the benefit of the minority that are at risk. The more realistic option available to physicians and to the state is voluntary patient compliance coupled with education and counselling.[41] Policies should be implemented to ensure women have access to prenatal care, are informed through counselling and classes and, where necessary, have access to drug rehabilitation programs.[40] With this background, pregnant women will be better prepared for possible outcomes and will comply more often with their physician's medical advice. These steps will encourage pregnant women to seek prenatal care and to provide accurate information without any fear of prosecution.

Developments in fetal therapy have enhanced the status of the fetus raising several legal and ethical issues. One of the most difficult of these issues is weighing the right of the mother to autonomy and bodily integrity against the right of the fetus to life and health. The pregnant woman has an ethical obligation to her fetus once it is viable to accept standard medical treatment for it. However, this obligation does not exist legally and should not, according to legal precedent, be enforced by the courts. Instead of trying to control the pregnant woman with laws and constraints, the government should ensure there is sufficient prenatal care for pregnant women, accompanied by support programs which provide education and counselling. Many more fetuses will benefit from this contribution than from forceful attempts to control pregnant women.

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Craniofacial Surgery: Correction of Orbital Hypertelorism

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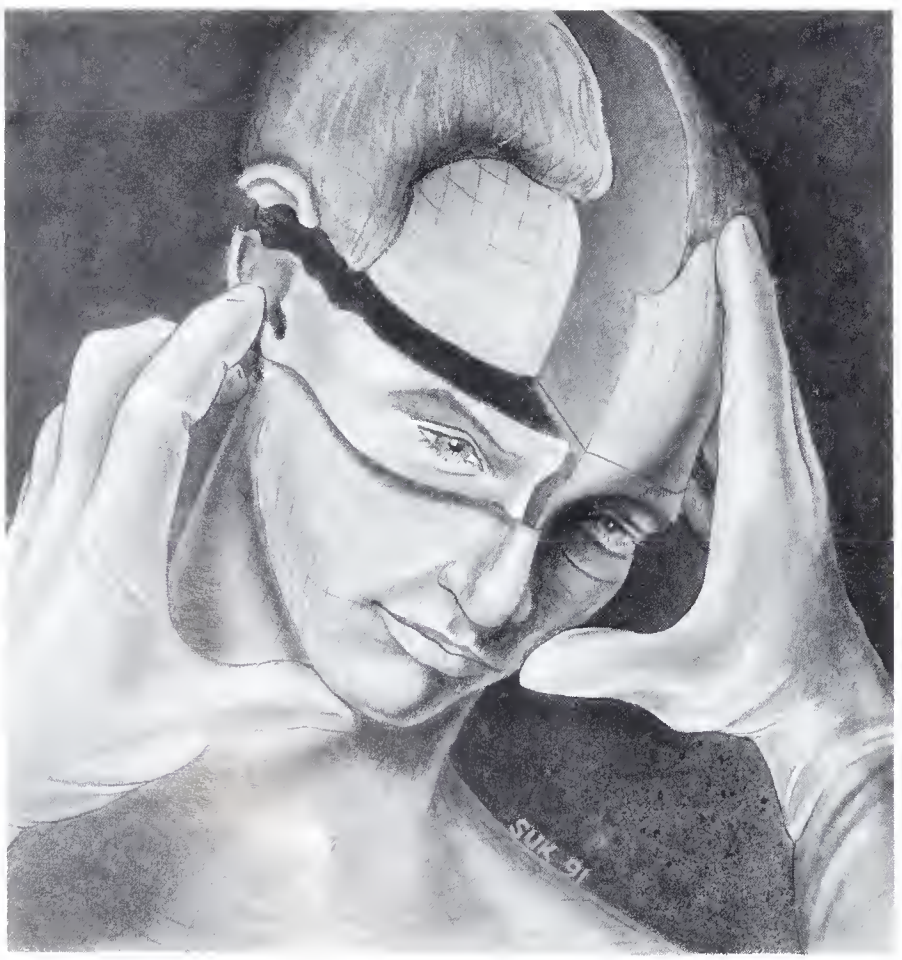
Abstract

Craniofacial surgery is a relatively new field in medicine which utilizes reconstructive surgical procedures to correct serious deformities involving the cranium, orbits and other facial bones. Orbital hypertelorism is a physical finding in which the transverse distance between the bony orbits is abnormally large. In this review, the etiologies of orbital hypertelorism are presented, several operative approaches for the correction of this deformity are described in detail, and operative complications are discussed. A case study is presented to demonstrate the quantitative and qualitative improvements in facial morphology and appearance possible with modern craniofacial reconstructive surgery. Careful preoperative planning, timing of surgery, and selection of a surgical procedure carried out by an experienced craniofacial team are crucial in the eventual outcome of the patient.



Introduction

Craniofacial surgery refers to surgical procedures requiring an intracranial approach used to correct complex congenital, neoplastic and acquired deformities involving the cranium, orbits, and other facial bones.[1] The birth of craniofacial surgery occurred in 1967 at the 4th Annual Congress of the International Confederation of Plastic Surgeons. It was here that Dr. Paul Tessier presented his work for the definitive treatment of craniofacial deformities, describing a radical and new approach which resulted in one of the major advances in modern reconstructive surgery.[1,2] By modifying osteotomies initially observed by Lefort in complex facial fractures, Tessier established three main principles: 1. Extensive areas of the craniofacial skeleton could be completely devascularized, osteotomized and repositioned and still survive. 2. The orbits could be circumferentially mobilized and repositioned without affecting vision. 3. The combined simultaneous intracranial and extracranial (subcranial) surgical approaches enable radical repositioning of structures and correction of major craniofacial deformities.[2,3] Prior to Tessier's work, complex craniofacial anomalies were often treated with superficial camouflage procedures, with only limited functional and aesthetic results.[2]

Craniofacial surgery has evolved over the past few decades, and specialized centres now care for the majority of patients with major craniofacial deformities. A team approach is required for the proper management of these patients. A typical team, as is found at the Hospital for Sick Children (HSC), is headed by a craniofacial plastic surgeon and includes a neuroradiologist, neurosurgeon, microsurgeon, anaesthetist, orthodontist, paediatric dentist, ophthalmologist, otolaryngologist, geneticist, paediatrician, physical anthropologist, psychiatrist, psychologist, social worker, nursing staff, physical and occupational therapist, audiologist and speech pathologist.[1,3]

Orbital Hypertelorism

Orbital hypertelorism (OHT) is a physical finding, not a syndrome, and is always secondary to another process.[1] Originally described by Greig in 1924 as "a great breadth between the eyes", OHT can be defined as abnormal spacing of the orbits with an increased interorbital distance.[3] OHT can

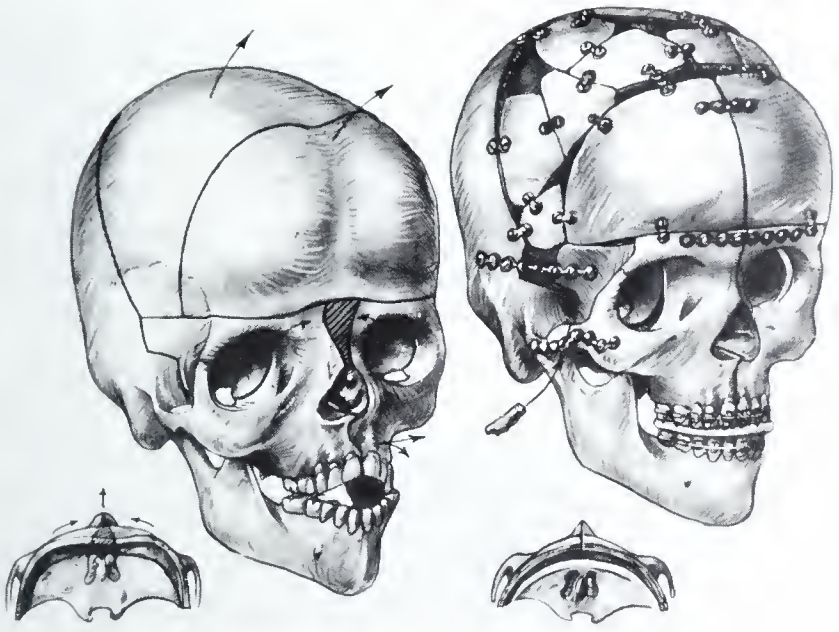


Figure 1a: Illustration of preoperative craniofacial morphology and planned and completed osteotomies with reshaping. Stabilization is achieved with cranial bone grafts and miniplate fixation.

be documented clinically by measuring intercanthal distance (the medial meeting point of the upper and lower lids) and interpupillary distance, or radiologically by measuring the interorbital distance on A-P skull X-rays or axial CT.[4] However, non-radiologic measurements may lead to misdiagnosis of OHT for pseudohypertelorism (or telecanthus), where there is lateral displacement of the medial canthi, occurring with epicanthal folds, blepharophimosis, Waardenburg syndrome or trauma.[1,4,5]

OHT is traditionally classified by the interorbital distance measured on an adult A-P skull X-ray, with 1st degree being 30-34 mm, 2nd degree 34-40 mm, and 3rd degree greater than 40 mm (normal range is 18.5-30.7 mm with a mean of 22.6 mm).[6] Severe OHT produces a marked facial deformity with an abnormal displeasing appearance.[15] The etiology of OHT can be divided into four main categories, major congenital facial clefting, craniosynostosis (premature closing of the skull and cranial base sutures), naso-frontal growths or neoplasms, and high energy facial trauma, as listed in Table 1.[3,5,6,8,9,10] The principal anatomic abnormality in OHT is transverse widening of the anterior ethmoid air cells, with a gradually increasing interorbital distance from the orbital apex to the rims. The sphenoid bone is usually normal and therefore the optic canals are in a normal position.[1,5] This anatomic arrangement allows the anterior half of each orbit, once osteotomized, to be brought closer together without impairing optical or oculomotor function.[1,5] OHT may be associated with conditions affecting the canthi, eye lids, globes, external ocular muscles, nasolacrimal system, and bones of the

Table I : Etiologies of Orbital Hypertelorism

Clefts:	Facial Clefts	– Cleft Palate – Unilateral Facial Cleft – Bilateral Facial Clefts – Median Facial Dysraphia – Bifid Nose – Proboscis Lateralis
	Cranial Clefts	– Unilateral Cranial Cleft – Bilateral Cranial Clefts – Dysraphias
	Combined Craniofacial Clefts	
Cranio-synostosis:	Isolated Craniosynotosis	– Unilateral Coronal Syntosis
	Craniofacial Dysostosis	– Apert Syndrome, Crouzon Syndrome, Kleeblattschadel (Coverleaf Skull Syndrome)
Tumours and Congenital Growths:	Frontal Encephalocele, Naso-Ethmoidal Encephalocele, Dermoid Cyst, Benign and Malignant Tumours	
Trauma:	High Energy Facial Smash with 3 or 4 Wall Orbital Fractures	

Figure 1b: Illustration of completed facial bipartition osteotomies. Tessier disimpaction forceps placed through the nose and mouth with pterygomaxillary spreader forceps placed through the coronal incision and infratemporal fossa to the pterygomaxillary suture regions for disimpaction of midface.

forehead, zygoma, nose and maxilla. When present, these associated abnormalities must be managed appropriately along with the OHT if the overall facial morphology is to be corrected.[11]

Preoperative Planning

In planning for the repair of OHT, input is gathered from appropriate members of the craniofacial team. Especially helpful are the clinical facial examination by the craniofacial surgeon, and the neuroradiologic examination, which may include A-P and lateral skull X-rays (cephalograms), and most importantly the axial, coronal, and 3-D CT scans.[1] Measurements are often performed on the patient anthropometrically, and on the cephalometric X-rays and CT scans, allowing for quantification of distances and volumes.[12] This assists in determining the extent of deformity and provides a realistic visualization of the craniofacial anatomy.[1,12,13] Preoperative orthodontic evaluation and treatment are often crucial to achieve a final reconstruction at the occlusal level. The specific reconstructive procedure and

Figure 2a: Preoperative frontal view.

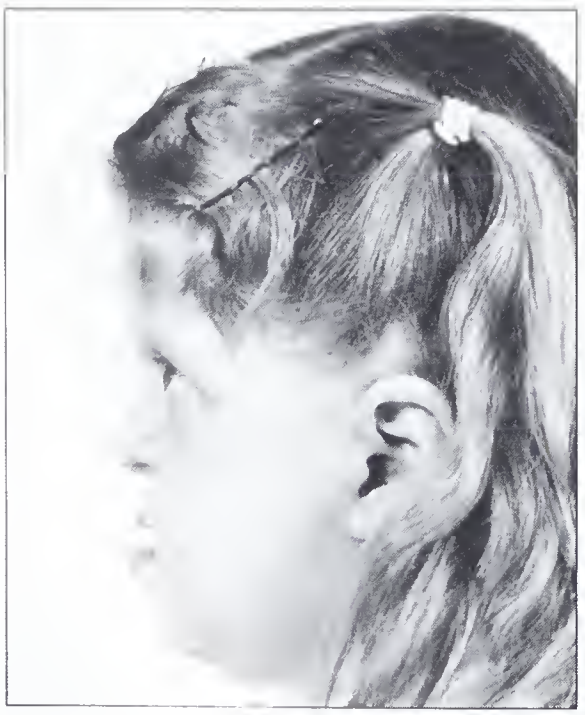


Figure 2b: Preoperative lateral view.

Figure 3a: Postoperative frontal view 1 year after reconstruction.

Figure 3b: Postoperative lateral view 1 year after reconstruction.

timing of repair are then selected, often with the need for a staged approach to facial reconstruction based on the residual growth potential anticipated in the cranio-maxillo-facial skeleton after initial reconstruction. When a non-progressive congenital anomaly is responsible for OHT, and if major functional problems (such as increased intracranial pressure, exophthalmos with corneal injury, amblyopia, or loss of binocular vision) are not present, the senior author prefers finalizing orbital reconstruction between the age of 5-7 years, at which time the cranio-orbital skeleton is more than 85% of the final adult size.[12] This coincides with the child entering grade school, when psychosocial issues begin to come to the forefront of the child's development.[12,13]

Operative Procedure

All procedures are performed under general anaesthesia with either oral or nasal endotracheal intubation and intensive intraoperative monitoring. The classic procedure for OHT reconstruction developed by Tessier and others relies upon circumferentially mobilizing and repositioning the orbits through a combined intracranial and extracranial approach.[1] Through a coronal scalp incision a neurosurgeon uses burr holes and a pneumatic saw to remove a segment of the frontal bone. The frontal and temporal lobes of the brain are then retracted posteriorly exposing the anterior cranial fossa and allowing for safe orbital osteotomies. The craniofacial plastic surgeon then further exposes the orbital region and using a subperiosteal dissection, frees the periorbital structures and orbital contents. The floor of each orbit and superior maxilla

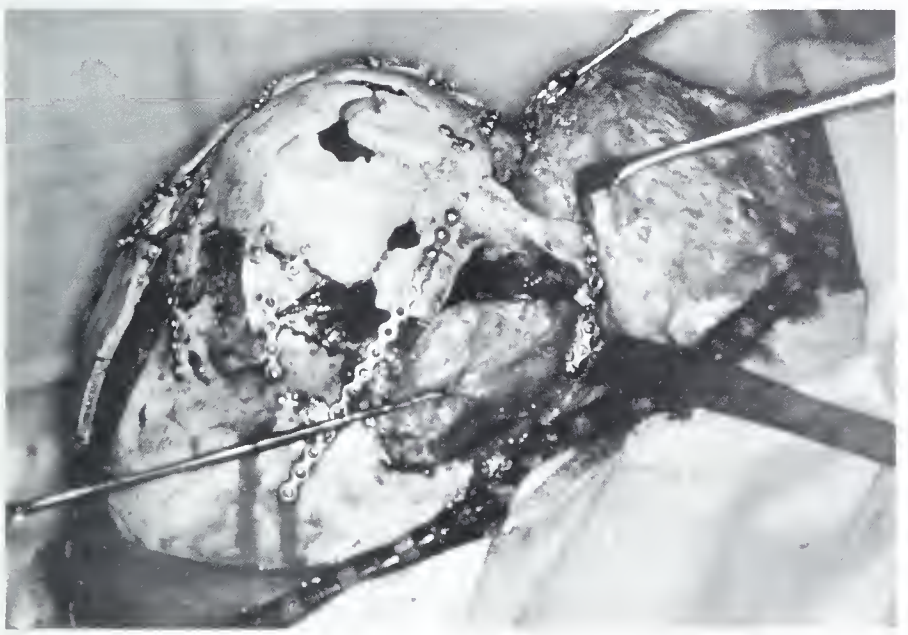


Figure 1c: Intraoperative lateral view of cranial vault, orbits, and zygomatic arches through coronal incision after osteotomies. Stabilization with cranial bone grafts and titanium miniplate fixation.

Figure 4: Three-dimensional CT scan reformations. Oblique view before and after reconstruction.

can be approached through either a subcilliary (the skin of the lower lid) or transconjunctival incision. Osteotomies are then performed using a pneumatic saw and osteotomes circumferentially in the orbital rims and all four orbital walls. Predetermined amounts of bone are removed from the interorbital region in the midline, and the enlarged ethmoid air cells are exenterated. The orbits are now freely mobile and are translocated medially to their new position. Internal fixation is obtained with metal bone plates and screws specifically designed for the facial bones, and with split or full thickness autogenous cranial bone grafts which are used to fill in the remaining gaps. Canthopexies and other secondary procedures such as nasal reconstruction are then carried out. All dural defects are repaired and the frontal bone is reshaped as required and replaced. The scalp flaps are closed in layers, generally over suction drains.[1,2,7,14]

The facial bipartition procedure (Figures 1a,b,c), initially described by Van Der Meulen, provides an alternative to the classic "total orbital osteotomies" approach.[15] This procedure has the advantage of addressing the other associated bony abnormalities which may be present in other regions of the craniofacial skeleton. Using a coronal incision and a combined intracranial and extracranial approach, the facial skeleton is split vertically in the midline from the frontal bone to the alveolar ridge of the maxilla. Osteotomies are performed horizontally along the supraorbital ridge and skull base anterior to the crista galli. The zygomatic arches are osteotomized and the pterygomaxillary sutures separated. Orbital osteotomies are then made through all four walls, the hard palate is split sagitally in the midline, and the bony nasal septum (vomer) is separated from the skull base (Figure 1a). The total midface and orbits are then mobilized using disimpaction forceps (Figure 1b). A triangular wedge of bone including frontal bone, ethmoid, nasal bones, septum and

maxillae is resected. The two facial segments are then repositioned allowing medial translocation of the orbits, nasal bones, and zygomas while simultaneously widening the maxillary arch width. Stabilization is obtained with multiple bone mini and microplates and screw fixation (Figure 1c). Intermaxillary fixation is then used to achieve and maintain a preferred occlusion of the jaws.[14,15,16,17] When indicated, this procedure offers certain benefits for improving facial morphology of the hyperteloric face. By mobilizing the two hemi faces "in toto", the interorbital, intertemporal, and interzygomatic distances can be diminished together with a widening and flattening of the dental arch and palate, which is often high arched ("keel" shaped) in OHT. The typically short anterior facial height is effectively lengthened. The midface, which is often retruded in craniosynostosis syndromes, can be advanced to increase facial and orbital depth and improve dental occlusion. Reshaping and repositioning of the frontal bone allows for expansion of the intracranial volume. This assists in reducing the increased intracranial pressure which is often associated with craniosynostosis (Figure 1a). The unerupted permanent maxillary dentition in younger patients is protected with the facial bipartition approach by avoiding osteotomies in the infraorbital region of the maxilla, which would be required with the standard "total orbital osteotomies" approach.[14,15,16,17]

A subcranial approach is a third option for correction of mild degrees of OHT or for pseudohypertelorism.[4] Through the incisions described above, a U-shaped osteotomy through the medial and lateral orbital walls and orbital floor is performed, leaving the orbital roof intact. Bone is resected from the nasal bones and/or the frontal processes of the maxillae, with exenteration of the ethmoid air cells. The orbits are mobilized, repositioned medially, and fixed with bone plates and screws. This procedure avoids the risks of an intracranial approach (such as brain damage) by remaining extracranial, but is often less effective in correcting the orbital deformity, and does not address associated deformities which may be present.[1,4,18]

Complications

Complications vary with the procedure, the age of the patient, the number of procedures previously carried out on the patient, and the experience of the craniofacial surgeon and team. Fortunately most serious complications, such as death and irreversible brain or eye damage, are rare and minimized when procedures are appropriately selected and then carried out by experienced craniofacial teams.[9,19] A multicentre retrospective report described complications from craniofacial surgery performed on 793 patients. The percentage of deaths was 1.6%, loss of visual acuity 0.3%, permanent brain damage 0%, infection including wound infection, osteomyelitis and meningitis 6.2%, and temporary CSF leak 1.1%.[1] This data, however, was collected from 1968-77 during the early formative years of craniofacial surgery. A more recent study from a series of 155 patients reported an infection rate of 1.3% and no cases of death, permanent brain damage or loss of vision.[9] Blood transfusions are often required, with one centre's average volume transfused being 660 cc per patient.[19] Other complications varying greatly with the specific procedure carried out are loss of olfaction, bone graft resorption and necrosis, and skeletal relapse.

Case Study

The patient presented is a five-year-old girl with Apert syndrome treated by the senior author. The patient was seen for evaluation by the craniofacial team at HSC at age 3. Her examination demonstrates OHT resulting from craniosynostosis. She has an abnormal cranial vault with brachycephaly, shallow orbits with proptosis, and midface hypoplasia with a class III anterior open bite dental malocclusion (the maxillary molars are too far posterior compared to the mandibular)(Figures 2a,b). Facial analysis was carried out with cephalometric X-rays, axial, coronal and 3-D CT, surface anthropometric measurements, and overall clinical impression. Quantitative measurements performed on CT scans reflected the clinical findings with interorbital distance of 30.0 mm (OHT), a cephalic length of 159.2 mm and cephalic width of 137.6 (brachycephaly), a globe protrusion of 21.1 mm (eye proptosis) and a zygomatic arch length of 36.8 mm (retruded midface)(the age matched normal means are 20.6 mm, 179.1 mm, 134.4 mm, 15.6 mm, 49.3 mm respectively).[12]

A modified facial bipartition osteotomy and cranial vault reshaping was carried out through an intracranial approach by the senior author in 1988 at HSC (Figures 1a,b,c). The patient experienced no post-operative complications and returned home approximately 2 weeks after surgery. Three years after the surgical procedure, significant improvement in her overall appearance remains, and a late post-operative craniofacial CT examination confirms that she is less hyperteloric with an interorbital distance of 23.6 mm, less proptotic with a globe protrusion of 13.1 mm, has better projection of her midface with a zygomatic arch length of 50.0 mm, has improved cranial vault shape and volume with a cephalic length of 176.3 mm and cephalic width of 134.7 mm, and improved dental occlusion (Figures 3a,b and 4).

Conclusions

The patient presented above illustrates the lasting improvement in appearance and function that is possible with meticulous pre-operative planning, careful timing of surgery, and thoughtful selection of an operative procedure to correct the abnormal craniofacial morphology. Functional, aesthetic, and psychosocial factors must all be carefully considered when evaluating a patient with a craniofacial deformity. Complex reconstructive procedures can be performed safely

on patients from 22 months of age onwards when carried out by an experienced craniofacial team.[19]

Acknowledgements

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Colorectal Carcinoma in a Young Female and Literature Review

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Abstract

Colorectal carcinoma is a common disease of later life which results in a high degree of morbidity and mortality. Through research, medicine has witnessed many advances in our knowledge concerning the pathogenesis of this disease. Despite our growing understanding of colorectal carcinoma, the knowledge gained over the past twenty years has had little impact on our ability to reduce mortality from this disease. This paper illustrates there is a need to always consider malignancy in the differential diagnosis of rectal bleeding, even in the absence of obvious risk factors or family history. The current literature concerning colorectal carcinoma is reviewed.

Case History

A previously healthy 28-year-old black Jamaican woman presented with rectal bleeding of several months duration which progressively worsened over the last three months. The bleeding accompanied bowel movements and was of sufficient quantity to tint the toilet water red, and leave a bright red stain on the toilet paper. Although she had been constipated since childhood, her bowel movements had now changed such that they were more frequent, of smaller volume, and accompanied by hypogastric cramps. There was no nausea, vomiting, or weight loss.

The patient possessed sickle trait. Otherwise the functional inquiry and past medical history were insignificant. She was divorced with two children, and had no family history of bowel pathology. She had never smoked.

Physical examination of the abdomen and rectum was unremarkable. The remainder of the examination was also normal.

Investigative Plan

In older age groups (>50 years), the most likely diagnosis for new-onset rectal bleeding and altered bowel habits is colorectal carcinoma.[1] In a 28-year-old woman, however, this disease is generally uncommon and other diseases must be considered in the differential diagnosis. Factors to consider include family history, associated diseases, and disease incidence. The initial working differential diagnosis for rectal bleeding based on this presentation is summarized in Table 1. Causes of upper GI and small intestine hemorrhage are very unlikely as they usually present with melena, or maroon stools, rather than frank red blood. In addition to the etiologies listed in Table 1, one must also consider extracolonic entities such as endometrioma.

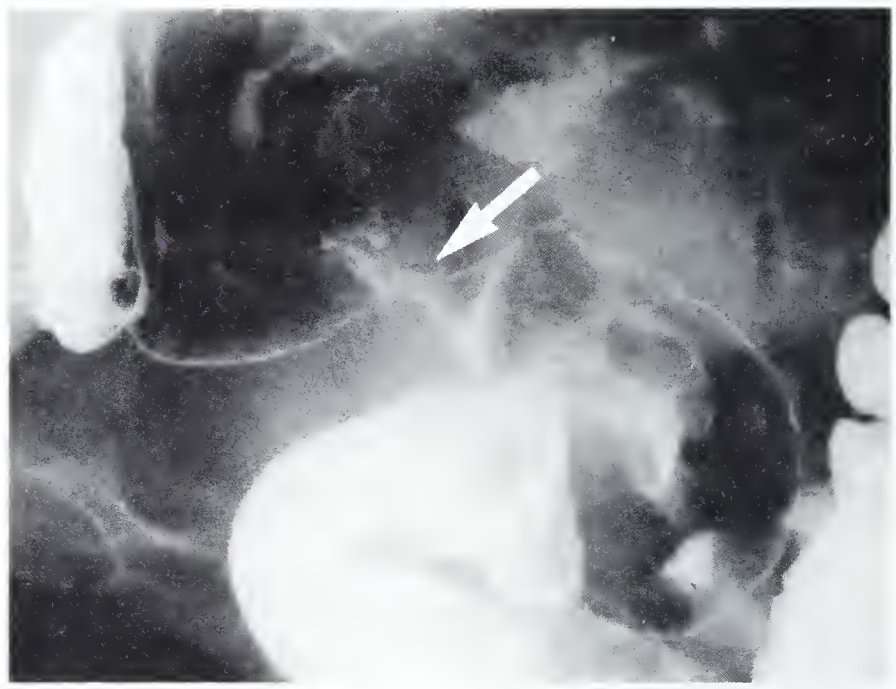

Figure 1: Investigative contrast barium enema clearly illustrates the marked narrowing and irregularity at the rectosigmoid junction. This filling defect, described as the "apple core" lesion, is characteristic for colorectal carcinoma.

Differential Diagnosis for Bright Red Blood per Rectum

1.	Hemorrhoids	
2.	Anal fissure	
3.	Rectal Trauma	
4.	Inflammatory Bowel Disease	- ulcerative colitis - Crohn's disease
5.	Diverticular disease	
6.	Colitis	- granulomatous - ischemic
7.	Ulcerating polyp	
8.	Colorectal carcinoma	
9.	Angiodysplasia	

Table 1: *The initial working differential diagnosis for a 28 year old woman of Jamaican origin with no family history or risk factors for colorectal cancer, presenting with bleeding per rectum.*

It is important to investigate the patient appropriately and efficiently to begin streamlining the differential diagnosis and arrive at the final diagnosis. Sigmoidoscopy is the simplest and most direct procedure to identify the majority of disorders causing the aforementioned symptoms and should be done initially. Sigmoidoscopy revealed the presence of a large, 5 cm diameter, ulcerating tumour 10 cm from the anal orifice. A biopsy of the lesion was sent to pathology. Additional investigations including routine blood chemistry, urinalysis, chest X-ray, and ultrasound of the liver, gallbladder, pancreas, spleen, and kidneys were normal.

Further investigation is required in the form of barium enema or colonoscopy to delineate the lesion and to identify any other synchronous lesions which may be present. This point is emphasized in that approximately 30% of patients demonstrate synchronous neoplasm(s), albeit usually benign.[1,2]

Figure 1 is the barium contrast enema obtained from this patient in the region of the rectosigmoid junction. Note the classic "apple core" lesion clearly illustrating the marked narrowing and irregularity of the lumen. This radiographic appearance is highly suggestive of malignancy. No other lesions were visualized.

The biopsy report confirmed the diagnosis of invasive adenocarcinoma. While carcinoma of the colon in a 28-year-old female can occur, it is very uncommon in this age group, especially in view of a negative family history for GI carcinoma or genetic syndromes such as familial polyposis, Gardner's syndrome, Turcot's syndrome, or nonpolyposis inherited colon cancer.[3-5]

Management

Surgery is the primary curative modality for patients with gastrointestinal carcinoma. Treatment consists of wide surgical resection of the lesion and its regional lymphatic drainage, even if distant metastasis has occurred since prevention of obstruction or bleeding may offer palliation for long periods. The abdomen is explored to determine resectability, and searched for metastases and synchronous lesions. Care is taken to prevent spread of malignant cells by avoiding unnecessary manipulation of the tumour.[6,7]

The choice of operation and extent of resection depends upon the location, configuration, and differentiation of the tumour, and the status of the patient. This patient received a low anterior resection with end-to-end anastamosis of the descending colon to the rectum, through a midline laparotomy incision. Prior to surgery she received a blood transfusion to improve her hemodynamic status.

At operation a 10 cm firm cicatrizing tumour was found with no evidence of peritoneal seeding. However, several palpable nodes along the inferior mesenteric artery (IMA)

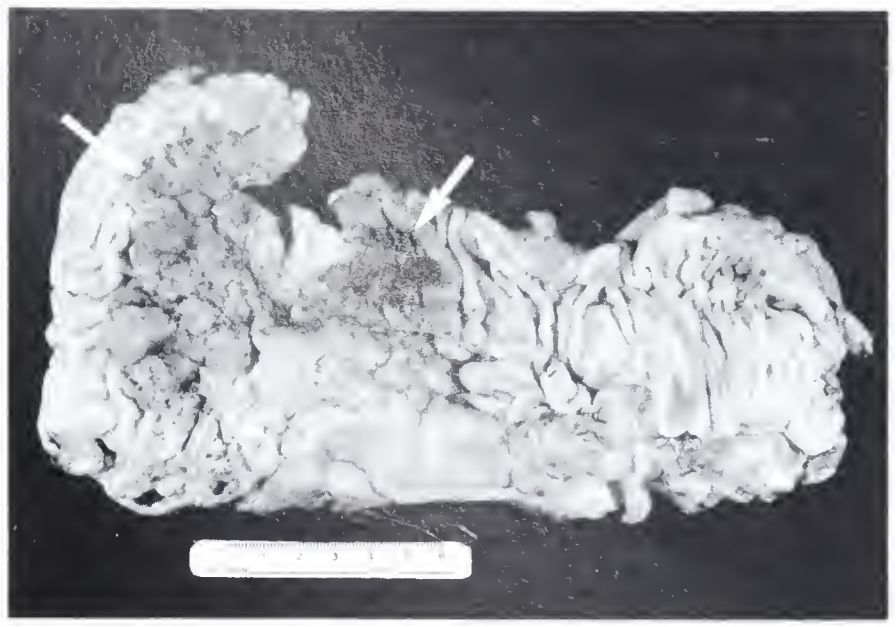

Figure 2: Gross characteristics of the resected tumour are shown. The mucosal aspect of the carcinoma exhibits abnormal morphology. Note the raised, rolled borders of this tumour.

distribution were found, the most proximal node being of 2 cm diameter at the origin of the IMA. The tumour, 3.0 cm of distal bowel, 9.5 cm of proximal bowel, its lymphatic distribution, and the IMA from its origin were resected. Figure 2 illustrates the gross characteristics of this tumour.

In view of the size of this tumour, and the young age of the patient, it was felt adjuvant pelvic radiotherapy would be required. In order to protect the normal small intestine from the effects of radiation, it was elected to fashion a sling out of the omentum to suspend the small intestine out of the pelvis.

The pathology report established a tumour with clear resection margins that had invaded through the serosa, but with no lymph node metastases. The patient was discharged two weeks following surgery with no postoperative complications. She was referred for follow-up radiotherapy, which is felt to be of benefit in reducing local recurrence.[8]

Follow-up and Prognosis

Follow-up is an essential component of management in the successful treatment of carcinoma. The goals are detection of recurrent, metastatic, or metachronous lesions, and are related to the location of the tumour, and the stage and grade of the disease.

Table 2 illustrates Dukes classification of cancer of the colon and rectum which correlates with five-year survival rates. This patient was classified as stage B2 which places her in a 60% five-year survival category.[9] Figure 3 is a diagrammatic representation of Dukes pathologic staging of colorectal carcinoma. Prognosis is also influenced by the grade of the tumour, that is, the degree of differentiation the tumour expresses. The microscopic findings of this tumour are shown in Figure 4.

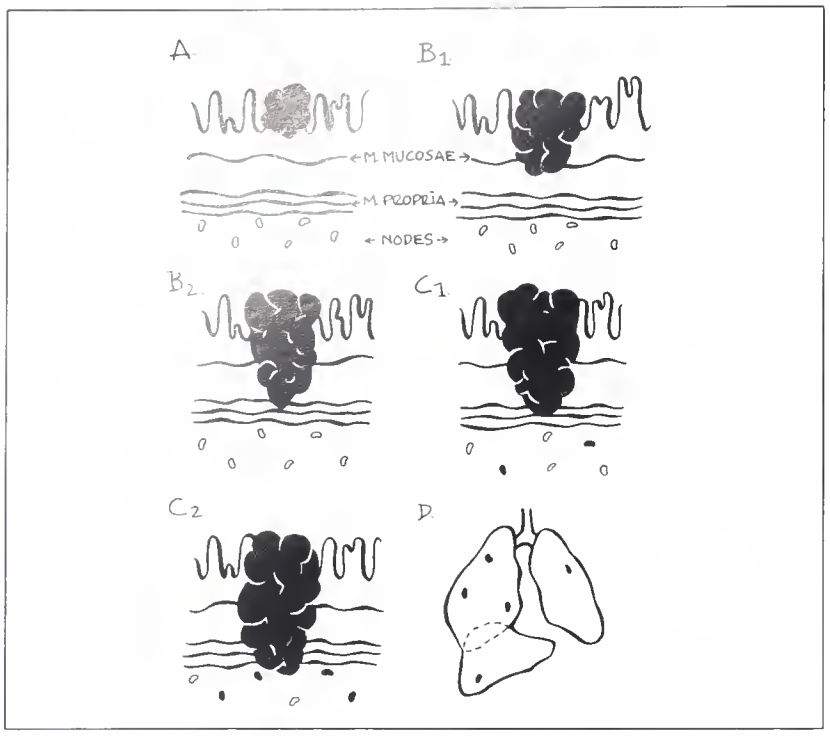

Figure 3: Diagrammatic representation of Dukes classification for colorectal cancer illustrates the degree of local tumour extention (stages A through C) and distant visceral metastases. Significant prognostic implications arise from this pathologic staging such that 5-year survival rates decline with greater tumour burden. (Modified from Robbins Pathologic Basis of Disease - special thanks to Kristi Vuorinen for the illustration)

Figure 4: Microscopic features of the tumour demonstrate adenocarcinoma morphology, which comprises 95% of colorectal cancers. The tumour is seen to penetrate into muscularis propria in this photomicrograph.

Follow-up in this patient will encompass several modalities. Carcinoembryonic antigen (CEA) levels will be measured every one to two months and will serve as a red flag should they begin to rise. Elevated CEA, although not specifically associated with colorectal carcinoma, is present in 70% of patients with cancer of the large intestine. Preoperative levels correlate with postoperative recurrence rate; progressive elevation during the follow-up period indicates recurrence is likely.[10]

Colonoscopy is performed one year postoperatively and then every two to three years if neoplasms are not seen in a thorough examination. It is important to search for and remove adenomas - tubular, villous, or tubulovillous pedunculated tissue projections (polyp) - as they may eventually give rise to adenocarcinoma.[3,11,12] Routine evaluation of the liver and chest is carried out to detect the presence of distant metastases. Barium contrast x-rays are not routinely done.[6]

Discussion of Colorectal Carcinoma

Cancer of the large intestine as a cause of death is second only to lung cancer in men, and breast cancer in women, among all cancers in adults. Despite the most recent advances in early

Dukes Classification

Stage	Description	5 Yr Survival
A	Tumour limited to mucosa	80 %
B1	Tumour through mucosa, still within bowel wall	60 %
B2	Tumour through entire bowel wall	
B3	Tumour adherent to or invading adjacent organs	
C1	Tumour limited to bowel wall, but nodes positive	30 %
C2	Tumour through entire bowel wall, nodes positive	
C3	Tumour adherent to or invading adjacent organs, nodes positive	
D	Distant metastases or unresectable tumour	5 %

Table 2: *Dukes classification for cancer of the colon and rectum is summarized. Note that more aggressive lesions correlate with reduced life expectancy. (Modified from Lange's Current Surgical Diagnosis and Treatment.)*

diagnosis, screening procedures, and identification of populations at risk, mortality from this disease has changed little over the past two decades. At present there is no established means of prevention, and there is no reliable and cost-effective means of screening to ensure early diagnosis.[1,13]

Colonic cancer is principally a disease of Western societies. Seventy-five per cent of cases are sporadic whereas 25% have an identifiable genetic association. Peak incidence occurs in the seventh decade of life; fewer than 20% of cases are diagnosed under 50 years of age. There is a male:female ratio of approximately 2:1 for tumours occurring in the rectosigmoid region, and no sex preponderance for tumours located proximally or in the rectum.[14,15]

The past decade has witnessed an increased incidence of colonic carcinoma in the right colon. Formerly, colonic cancer was much more commonly located in the colorectal region. It has been proposed this altered pattern is due to the increased use of flexible sigmoidoscopy and removal of adenomatous polyps, resulting in reduced incidence of rectosigmoid carcinoma.[16,17]

Much of the diagnosis relies upon the presentation, and this differs according to the location of the lesion. From the view point of morphology and clinical behavior three distinct presentations exist and are summarized in Table 3. Figure 5 shows the relative distribution of cancer as it occurs in the large intestine.

Clinical presentation according to location of colon cancer

Right Colon:	1. Unexplained weakness or anemia 2. Occult blood in feces 3. Dyspeptic symptoms 4. Persistent right abdominal discomfort 5. Palpable abdominal mass
Left Colon:	1. Change in bowel habits 2. Gross blood in stool 3. Obstructive symptomatology
Rectum:	1. Rectal bleeding 2. Change in bowel habits 3. Sensation of incomplete evacuation

Table 3: *Tumours in different locations of the large intestine manifest as different clinical presentations. The associated findings are summarized according to location of the pathology.*

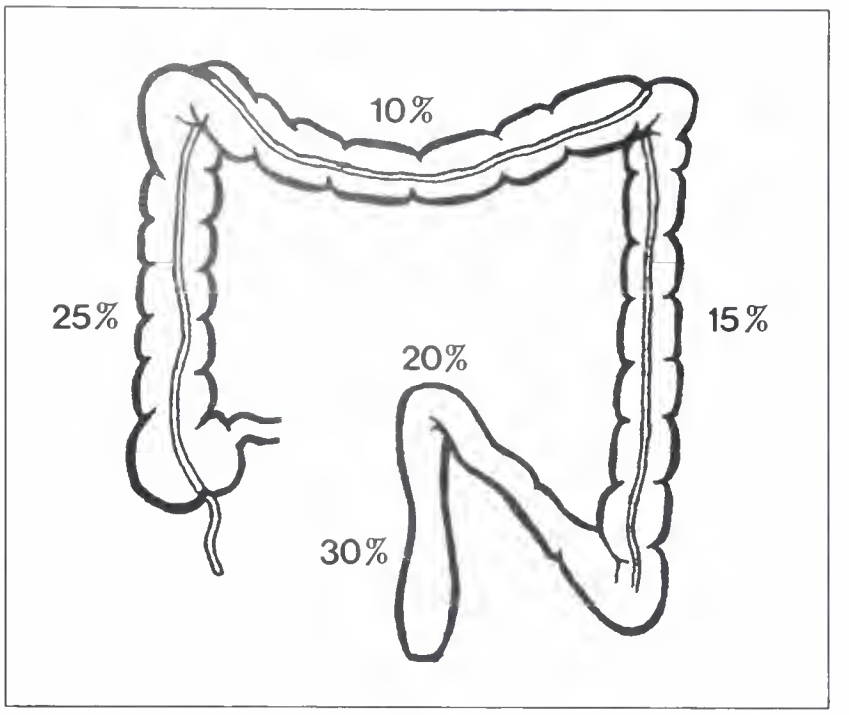

Figure 5: The distribution of incidence of carcinoma within the large intestine is illustrated. (Modified from Lange's Current Surgical Diagnosis and Treatment)

In terms of pathology, ninety-five per cent of colorectal cancer is adenocarcinoma. Right-sided carcinomas usually begin as sessile lesions that progressively assume a polypoid, fungating appearance to become bulky, cauliflower-like lesions. As they often occur in the capacious cecum where the contents are liquid, they do not lead to obstruction until very late and are silent for a long duration, thus accounting for their frequent presentation as iron deficiency anemia. Left-sided carcinomas are typically flat, invasive lesions that encircle and encroach the bowel lumen leading to obstruction of the solid contents. Eventual ulceration of the lesion produces frank bleeding.[12,15]

Eighty per cent of diagnoses are made at a stage when all apparent diseased tissue can be surgically removed. Incurability is likely due to occult residual cancer from micrometastases. The patient discussed will receive adjuvant radiotherapy as there is a significant risk of local recurrence for rectal carcinoma (20 - 50%) if the tumour penetrates through the rectal wall or if lymph nodes are involved. This modality is used to extend the definitive therapy beyond the limits of surgical excision. Postoperative radiotherapy for stages B2, C1, and C2 carcinoma of the rectosigmoid region reduces the incidence of local recurrence and may improve survival rates.[6,8]

Recent studies have shown the value of chemotherapy as a treatment for metastasis or recurrence of colonic and rectal cancer. The current mainstay of therapy for metastatic or recurrent carcinoma is 5-fluorouracil and folinic acid.[18] In addition, adjuvant therapy with levamisole and 5-fluorouracil is standard postoperative treatment for Stage C colonic cancer.[19]

Although the etiology of colorectal carcinoma is still obscure, abundant clinical and histopathological data suggest most, if not all, colorectal carcinomas arise from preexisting adenomas. Both environmental and hereditary factors contribute to the development of colorectal neoplasia.

Environmental Factors

The high incidence of colorectal cancer in Western societies has directed the attention of researchers to the role that diet may play in its genesis. Studies have demonstrated that immigrants from Japan, where the incidence of colorectal cancer is low, assume the increased North American incidence after the first generation.[20]

It has been proposed that diets low in fibre and high in saturated fats and refined carbohydrates may predispose to colorectal cancer. Another theoretical mechanism is that a low fibre diet results in low fecal bulk which reduces transit time and increases the concentration of bile acids which contact bowel mucosa, thereby increasing the opportunity for interaction between carcinogens and cells susceptible to neoplastic change.[21]

More likely is the fact the above dietary composition alters the colonic flora such that tumour promotor concentration is elevated through secondary metabolism of the increased cholesterol and bile acids which accompany a high fat diet. Breakdown of these bile acids and sterols creates a procarcinogen that acts as a proliferator of cancer in susceptible cells, that is, cells already containing altered DNA. Additionally, fibre, particularly soluble fibre, increases colonic fermentation and through this process may protect the colonic epithelium.[13, 21-24]

Genetics of Colorectal Cancer

Colorectal cancer genesis is a multistage progression that involves the necessary steps of initiation, promotion, and proliferation. A large body of literature demonstrates the role genetic mutations play in tumourigenesis.[3-5,7,14,15,25-28]

It appears specific alterations of particular genes are prerequisite for the initiation of colorectal cancer. These tumours arise through mutations in two classes of genes: oncogenes which act in a positive fashion for neoplastic transformation when activated, and suppressor genes which lose their tumour-suppressing ability when mutated. Inactivation of tumour suppressor genes appears to predominate.[25]

Molecular and histopathological studies demonstrate colonic neoplasms progress along a continuum beginning with normal mucosa, and terminating at frank malignancy. This model describes a successive gradation of changes involving the progression through benign stages. Polyps are benign neoplasms that develop following genetic mutations, and have been shown to be precursors for malignancy under certain conditions. This is no more evident than in familial polyposis coli, a disease in which polyps coat the colonic lining, where malignancy eventually develops in 100% of affected individuals.[14,26-28]

Genetic alterations shown to be necessary for tumourigenesis include the activation of the ras-oncogene, coupled with allelic deletion of tumour suppressor genes located on chromosomes 5, 17, and 18. The total accumulation of genetic changes, rather than their order with respect to one another, appears to be responsible for determining the tumour's biologic properties. At least four to five gene mutations are required for the formation of a malignant tumour; fewer mutations are seen in benign neoplasms.[25]

Conclusions

Although our understanding of colorectal carcinoma has increased markedly during the past twenty years, morbidity and mortality from this disease remains virtually unchanged. For this reason it is paramount that the medical profession educate the general public regarding colorectal cancer, and the often vague symptomatology it may produce. Very often patients ignore these early symptoms, and indeed, these symptoms may be insufficiently investigated by physicians. It is recommended that all patients over 45 years of age receive occult blood screening annually. Additionally, high risk patients (i.e. family history or previous cancer) should receive regular sigmoidoscopy and colonoscopy, with removal of suspicious polyps.

The case presented clearly illustrates the need to always consider carcinoma even in the younger population, and in the absence of any risk factors. Early detection and prompt treatment are paramount for the best possible outcome in a patient with colorectal carcinoma.

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Case Report: The Other Side of Skin Cancer

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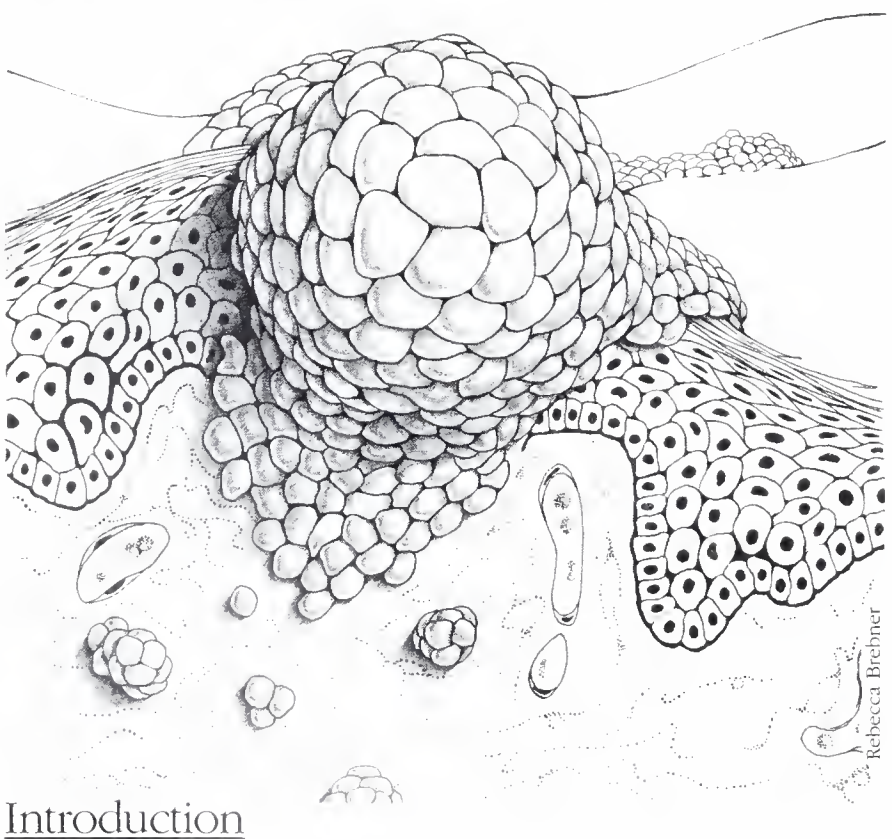
Abstract

Skin cancer is commonly a localized disease. Approximately 95% of cases can be cured with early local treatment. Melanoma poses a more serious threat, as metastases with this skin cancer are more common, and prognosis is dependent upon the depth of invasion.

This paper presents a case history with a brief discussion on a spindle cell variant of malignant melanoma and problems with diagnosing its cell of origin. As well, it reveals the other side of skin cancer, that is, the possibility of extreme aggressiveness with a rapid lethal outcome.



Introduction

This paper has two objectives: to demonstrate the possible aggressiveness of some skin cancers, and to illustrate the diagnostic difficulties associated with malignant spindle cell tumors of skin.

Case Report

A 66-year-old woman with a long history of a flat brown lesion on her left mid-upper brow, presented with the recent development of a firm, raised, flesh-colored area in its centre.

The lesion was excised with reportedly clear margins. It recurred one year later and was again excised, however, the margins were not clear. Four months later the patient presented to another hospital with the lesion having recurred. It was subsequently removed (margins clear). Two weeks later the patient complained of a rapid development of severe left facial pain and weakness. On examination, a left facial palsy accompanied by left parotid gland swelling was noted. A parotid gland biopsy was carried out. Subsequently, the patient underwent a left parotid gland excision and upper neck dissection with the sacrifice and reconstruction of the left facial nerve. Unfortunately, the tumor continued its aggressive course, with bony metastases being identified in the skull (Figure 1) and left femur. The patient was referred to Princess Margaret Hospital for consideration of combination chemo- and radiotherapy. Soon after her admission the patient succumbed to her disease.

Pathology

Materials and Methods: The third excisional biopsy from the forehead lesion, left parotid gland biopsy and the subsequent parotidectomy specimens were fixed in phosphate buffered 10% formaldehyde fixative, processed in the usual manner and stained with Hematoxylin & Eosin (H&E) for light microscopic examination.

Immunohistochemical staining for desmin, actin, S-100, and melanoma associated antigen (see Table 1 for tumor specificity[1,2,3,4]) was performed on tissues from the third skin biopsy, parotid biopsy and parotidectomy specimens by the Avidin-Biotin Complex (ABC) technique as described by Hsu et al.[5]

A sample of the skin tumor was fixed in 2.5% glutaraldehyde, processed in the usual manner and examined under the electron microscope. Histologic sections of the first two skin excisional biopsies from another institution were reviewed.

Description: The third excisional biopsy from the forehead lesion, left parotid gland biopsy, and left parotidectomy specimen all show a malignant spindle cell tumor of similar morphology (Figure 2A). The first two skin biopsies were reviewed. A malignant spindle cell tumor of similar morphology was present in the subcutaneous tissue, however, the first biopsy also showed a malignant melanoma present in the epidermis (Figure 2B).

Figure 1: A right lateral skull x-ray showing lytic metastatic bony lesions within the mid-parietal bone region.

Immunopositivity was noted for desmin, actin and melanoma associated antigen, but negative for S-100 in the final immunomarker assessment (Table 2 and Figures 3a/b). Electron microscopic examination was performed on the parotid gland biopsy and the tumor cells did not show smooth muscle differentiation. Premelanosomes and melanosomes were not observed.

Discussion

It appeared from both the clinical behavior and the difficulty making a histopathological diagnosis that this tumor represented a rare melanoma with malignant spindle cell characteristics.

Of the rare malignant spindle cell tumors of skin, the differential diagnosis would include spindle cell carcinoma (squamous cell variant), spindle cell sarcoma (leiomyo- and fibrosarcoma, malignant fibrous histiocytoma), and malignant melanoma (spindle cell variant). Since it is extremely difficult to identify the cell of origin of these tumors by light microscopy alone[6], immunomarkers (Table 1), and EM analysis, were utilized.

At one point, a tentative diagnosis of leiomyosarcoma was arrived at in light of the immunomarker stain results (desmin positive, S-100 negative, see Tables 1 and 2). The clinical behavior of the tumor in this case is unlike that of cutaneous leiomyosarcoma. Dahl et al[7] reports them to be extremely rare, occuring most frequently in the 60 to 70-year-old age group (two times more common in males), and most commonly located on the extremities. They should be treated by wide surgical excision. Local recurrences are seen commonly. Metastases occur less often and only after a

Table 1 : Immunomarkers utilized in case work-up

Antibody	Specificity For Cells In Skin	Tumor Types Associated With Positive Staining
S-100	melanocytes, Langerhans neural tissue	neuronal tumors, malignant melanoma, desmoplastic malignant melanoma
Desmin **	muscle cells	rhabdomyosarcoma, leiosyosarcoma
Melanoma associated antigen	malignant melanocytes	malignant melanoma
Actin	muscle cells, mycepithelial cells, myofibroblasts, neo plastic cells with contractile differentiation	leiomyoma, leiomyosarcoma, rhabdomyosarcoma

** = a nervous-system specific cytoplasmic protein (where the origin of melanocytes is via neural crest cells)*
*** = an intermediate filament of normal skelatal and smooth muscle, demonstrated in some smooth muscle tumors*

Table 2 : Summary of immunomarkers and pathology diagnosis

Tissue	Immunomarkers				Diagnosis
	S-100	Desmin	Actin	Melanoma Associated Antigen	
1st Ex Bx	-	-	-	-	IMM
2nd Ex Bx	-	-	-	-	LYM
3rd Ex Bx	neg	pos	pos	-	LYM
Parotid Bx*	neg	neg	pos	-	M-LYM
Parotid Bx* *Repeat Stain*	-	-	-	pos	
Parotidectomy	neg	neg	pos	-	M-LYM
Parotidectomy *Repeat Stain*	-	-	-	pos	M-MM

Bx = Biopsy, Ex Bx = Excisional Biopsy, (-) = not done, IMM = Invasive malignant melanoma, LYM = leiomyosarcoma, M-LYM - metastatic leiomyosarcoma, M-MM = metastatic malignant melanoma
** = EM performed (see Pathology)*

prolonged period of time usually in patients with large tumors or tumors showing a high mitotic rate. Electron microscopy, which is especially helpful in the diagnosis of smooth muscle tumors[8], showed no evidence of smooth muscle differentiation in this particular case. This, combined with inconsistencies of the patient's history when compared to the natural history of the disease, forced a reconsideration of this diagnosis.

Malignant melanoma was then felt to be more likely, despite S-100 negativity (the S-100 protein, although a good marker for malignant melanoma, is not specific for malignant melanocytes[9,10], nor does its absence entirely exclude this diagnosis). Electron microscopy also revealed the absence of both premelanosomes and melanosomes. Staining, however, with monoclonal antibody to melanoma associated antigen

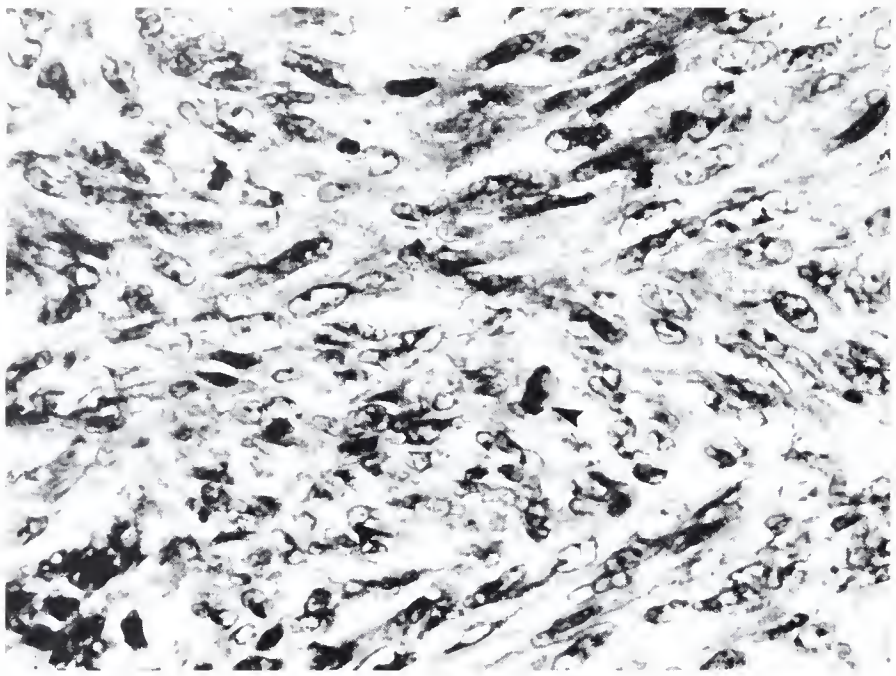


Figure 2a: Malignant spindle cell tumor in subcutaneous tissue (of third Ex Bx) with pleomorphic hyperchromatic nuclei and mitotic figures (arrowhead). H&E. Original magnification 400X.

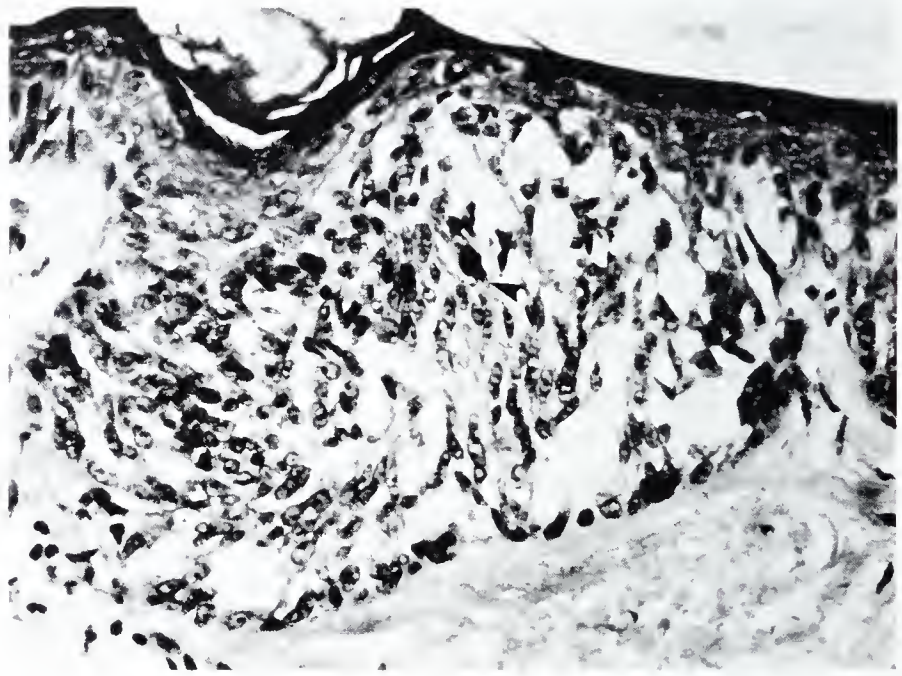


Figure 2b: Malignant melanoma in epidermis (first Ex Bx). Mitotic figure noted (arrowhead). H&E. Original magnification 250X.

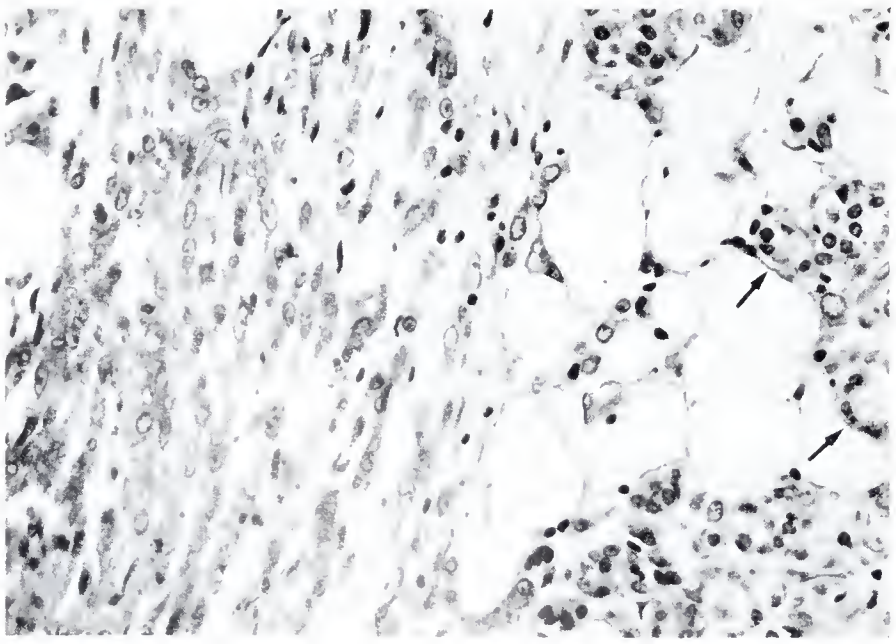


Figure 3a: Malignant spindle cells in parotid gland: melanoma associated antigen marker. Cytoplasm of malignant spindle cells staining diffusely (left half of field). Ductular cells of parotid gland are negative (arrow). Hematoxylin counterstained. Original magnification 250X.

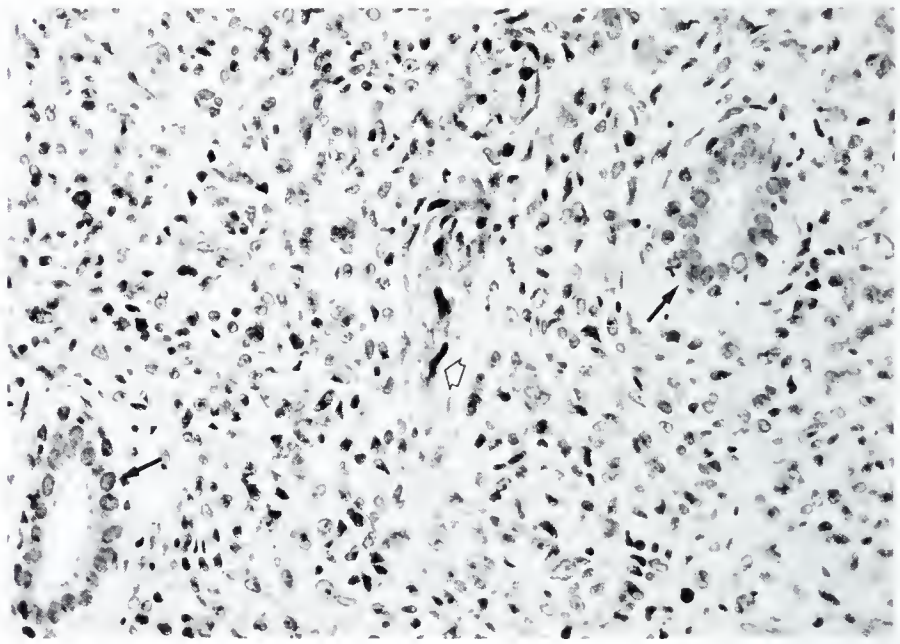


Figure 3b: Malignant spindle cells in parotid gland: S-100 marker. Normal neural tissue is positive (open arrow). Malignant spindle cells and ductular cells (arrow) of parotid gland are negative. Hematoxylin counterstained. Original magnification 250X.

was positive. This indicates, therefore, that the tumor is likely an amelanotic melanoma with malignant spindle cell characteristics.

The immunopositivity with desmin and actin might represent a cross-reactivity of the tumor cell antigens.

Switching briefly to consider the astounding aggressiveness of this tumor, which claimed the patient's life within nine months of its initial recurrence, one may question if an earlier and correct diagnosis would have changed the overall outcome in this case. In light of what was done for this patient from initial intervention onward, it seems unlikely the outcome would have changed (total surgical removal of the primary lesion with subsequent neck dissection were the principle modalities required for control of this patient's disease). Management of metastatic disease in malignant melanoma is very difficult as chemo- and immunotherapeutic modalities are ineffective in the majority of patients. It is not fully understood why similar lesions are aggressive in some individuals while in others they are not.

Summary

This case report demonstrates the difficulties in arriving at a pathological diagnosis of a malignant spindle cell tumor of skin. The diagnosis of amelanotic melanoma with malignant spindle cell characteristics, was finally arrived at, but not before the diagnosis of leiomyosarcoma was ruled out through EM and specific immunomarkers, namely, monoclonal antibody to melanoma associated antigen. This case also demonstrated both the frightening ability of some skin cancers to run a rapid, relentless course, and the almost futile attempts (in retrospect) to arrest its progression..

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Diagnosis and Management of Thumb Carpometcarpal Joint Osteoarthritis

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Abstract

The wide range of motion of the thumb is due to the unusual nature of its carpometacarpal (CMC) joint. Osteoarthritis is common in this joint, probably because of its high degree of use in load bearing function. The purpose of this article is to describe the radiographic stages of osteoarthritis, relate these to clinical appearance, and discuss treatment modalities for each stage of disease.

Illustrative Case Report

An active 70-year-old left hand dominant woman was referred to the hand clinic by the rheumatology service. She had a seven year history of left thumb base pain which had become refractory to treatment with intermittent intra-articular steroid injections and non-steroidal anti-inflammatory drugs (NSAIDs). Her present complaints included morning stiffness and ache which had progressed more rapidly during the previous eighteen months. Her activities were curtailed; she could not write comfortably and had difficulty buttoning her clothes. She felt that her left hand had become relatively useless. Her medical history was otherwise non-contributory.

Physical examination revealed dorsal subluxation of the left thumb carpometacarpal (CMC) joint and pain throughout the range of motion. There was slight ulnar deviation of the fingers. Lateral thumb radiographs (Figure 1) revealed bilateral Stage III degenerative changes in the CMC joint. A diagnosis of left thumb CMC joint osteoarthritis was made.

She was admitted to hospital for flexor carpi radialis tendon interposition arthroplasty and ligament reconstruction. At the time of surgery, inspection of the joint revealed total loss of articular cartilage, subchondral sclerosis, and osteophytes. The post-operative radiograph showed joint space widening at the site of joint debridement. A cast was worn for two weeks, followed by splinting for three additional weeks. Post-operatively there was full painless function as well as reduction of the subluxation of the joint.

Introduction

Osteoarthritis (OA), with loss of articular cartilage, and reactive changes in marginal and subchondral bone[1,2], is the most common form of debilitating joint disease in society. Osteoarthritis commonly affects the basal joint of the thumb[3], and has a major effect on hand function because of this joint's uniquely important role. The diagnosis and treatment of this common problem is presented.

The unique basal joint of the thumb allows prehensile activities and bears half the load in these functions[4]. The CMC joint is composed of two perpendicularly arranged saddles and has the widest range of motion of any joint in the hand[4-9]. The joint surfaces are in complete contact ("congruous") during abduction-adduction and flexion-extension[4-6]. For

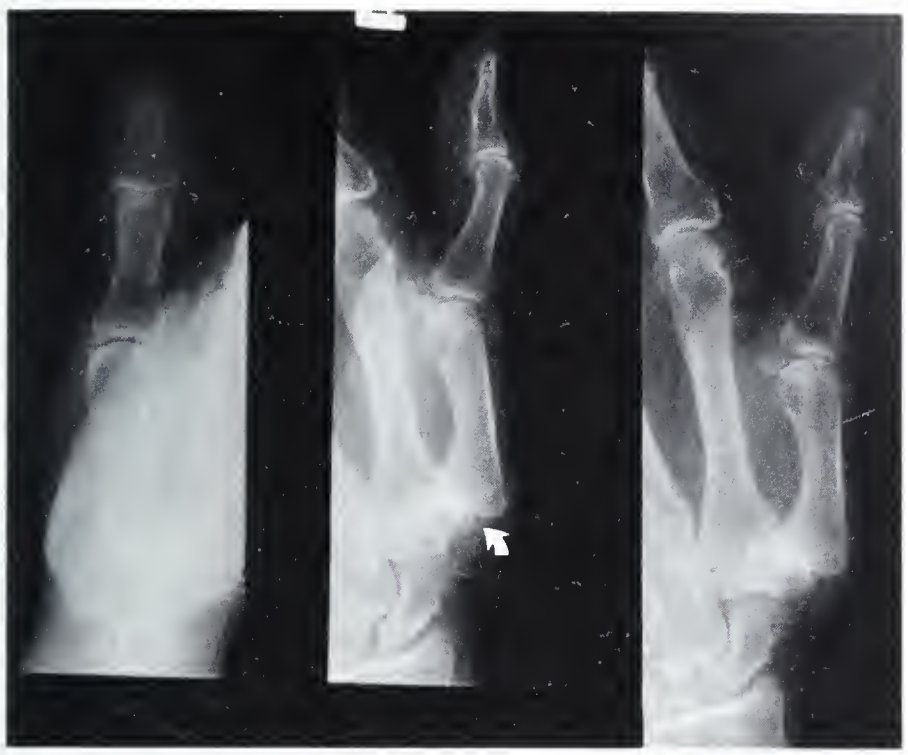

Figure 1: A lateral thumb radiograph of the patient showing Stage III articular changes in the left TM joint (arrow). There is joint space narrowing, more than one third subluxation, and osteophytes larger than 2 millimeters in diameter.

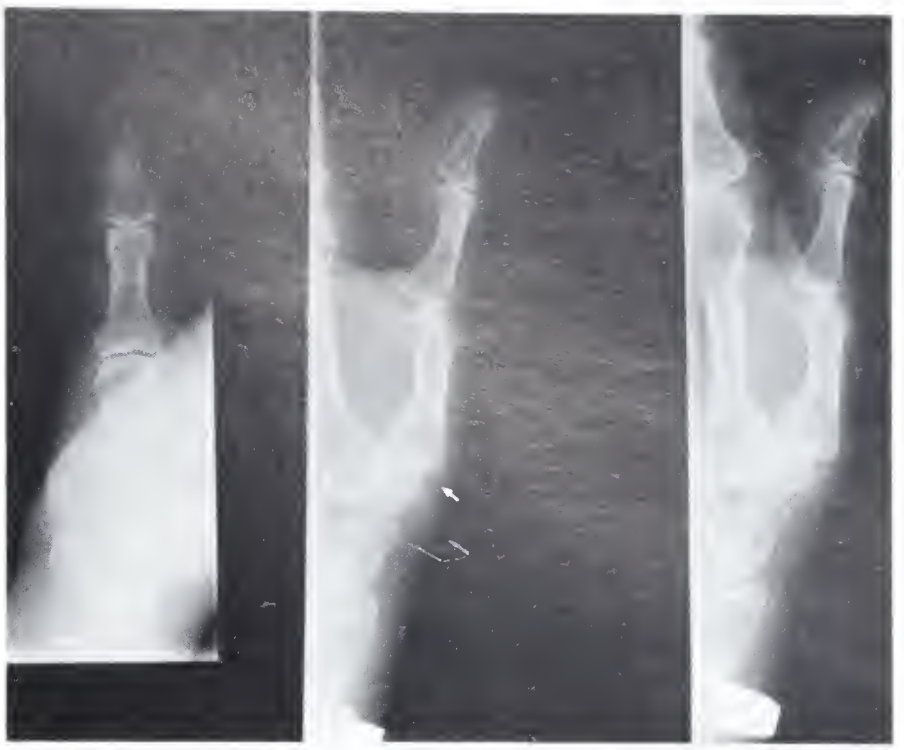

Figure 2: Stage I radiographic degenerative changes of the thumb CMC joint (arrow). The joint shows slight joint space narrowing, normal articular contours, and very little subluxation.

opposition and circumduction, the surfaces of the metacarpal and trapezium twist into an incongruous relationship, which is intrinsically less stable than the congruous position. In the twisted position, the ligaments are pulled tight and stabilize the joint. One or two millimeters of laxity of these ligaments accentuates the joint surface incongruity, causing abnormal pressure patterns which can result in degenerative changes in the articular cartilage[4-6].

The etiology of ligament hypermobility and subsequent cartilage degeneration is largely unknown. Occasionally, trauma or inflammatory conditions such as rheumatoid arthritis are responsible[5,10]. Idiopathic arthritis, the most common type, frequently occurs in postmenopausal females with a combination of a shallow trapezium concavity[11] and decreased cartilage repair[1].

In osteoarthritis, cartilage undergoes damage, allowing an ingress of water with hydrophilic swelling of proteoglycans. Microscopic changes include fibrillation of the superficial layers of cartilage which may progress to focal erosions and eventually total loss of cartilage. Subchondral bone thickens in response to the load, and bone remodels to redistribute this load, resulting in osteophyte formation. Some degree of synovitis may also be present, secondary to the inflammatory reaction to joint debris[2,3].

Presentation of Osteoarthritis

OA presents typically with pain and stiffness of the affected joints with use and occasionally there may also be short-lived morning stiffness[3,12]. Physical examination often shows joint enlargement, deformity (squaring), crepitus, and limitation of range of motion[3,12]. Onset of symptoms is insidious and prevalence increases with age[2,3].

Radiographic degenerative changes of the thumb CMC joint have been grouped into stages (Table 1) but these only correlate loosely with the anatomical and clinical aspects[5,13].

Radiographic Stage I (Figure 2), the "synovitis" stage, is characterized by slight widening of the joint space with normal articular contours and less than one third subluxation. Clinical correlates include effusion, hypermobility, and normal articular surfaces upon direct inspection[14].

In Stage II (Figure 3), the CMC joint is at least one third subluxated with osteophytes of less than two millimeters diameter. Although there is significant capsular laxity, clinically there is less hypermobility than in Stage I, and minimal crepitus[5].

Stage III (Figure 1) shows slight joint space narrowing, more than one third subluxation and fragments larger than two millimeters in diameter. Subchondral sclerosis may also be present[14]. Of note is that the scaphotrapezial (ST) joint is unaffected[14]. Crepitus, stiffness, and tenderness localized at the palmoradial trapeziometacarpal (TM) joint line are apparent.[15]. Direct inspection of the joint reveals that less than one third of Stage III joints have anatomical changes[13].

In Stage IV (Figure 4) radiographic disease, there are advanced degenerative changes at all facets of the trapezium. There is

Summary of Stages of Radiographic Degenerative Changes of Articular Surfaces in Osteoarthritis of the Basal Joint of the Thumb

Stage	Characteristics
I	Slight widening of joint space Articular surfaces normal Less than one third subluxation
II	Calcific fragments less than 2 mm diameter At least one third subluxation
III	Slight narrowing of joint space Calcific fragments larger than 2 mm diameter Greater than one third subluxation
IV	Vary narrowed joint space Large calcific fragments Major subluxation Erosion of dorsoradial facet of trapezium

major subluxation of the metacarpal, a very narrowed joint space, large osteophytes, and erosion of the dorsoradial facet of the trapezium[5]. Clinically, the metacarpal base is dorsally subluxed[5]. As in Stage III, pain and tenderness are principal features but are found over both the ST and TM joint lines[15]. Anatomically there are never more than two degenerated facets; half those with TM pathology also have ST involvement[13].

Clinical Assessment

Since radiographic changes do not correlate well with anatomical deformity, and no known therapy alters the usually benign natural history of the disease, treatment of OA of the thumb CMC joint is based mainly on symptomatology[3,13,14]. Localizing the patient's complaint of pain to the thumb CMC joint is important. Point tenderness as described above is common. Finkelstein's test, where the hand is forcefully ulnar deviated, produces pain in deQuervain's tenosynovitis but this is usually absent in OA[15]. Articular degeneration will produce pain when the articular surfaces are compressed together and rotated (the "grind test")[15,17]. Advanced disease produces pain on passive movement, which is relieved by instilling local anesthetic into the TM joint space[15]. Deformity must be assessed prior to treatment planning. The classic deformity is CMC flexion with subluxation and metacarpophalangeal hyperextension[15].

Treatment

Conservative measures are used to manage early disease, including rest from aggravating manual tasks, functional splints to allow joint rest, warm soaks, and medications[2]. In episodic disease, analgesics suffice; otherwise a 4-6 week trial of NSAIDs (if there are no contraindications) may be necessary. All NSAIDS are thought to be equally effective[12,18].

Intra-articular steroid injections may be used for acute flare-ups of single joints[19], and may be repeated every six months[12,18] for several years. After the discomfort of injection resolves, relief can last months[20]. Steroid injections are ineffective if articular cartilage is severely denuded, and cannot be repeated indefinitely[16].

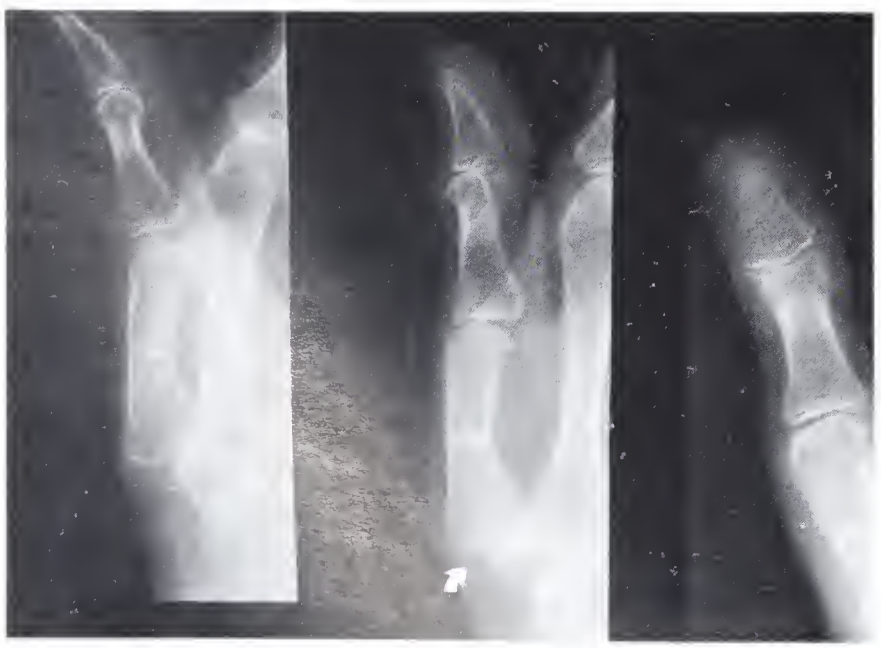

Figure 3: Stage II radiographic degenerative changes of the thumb CMC joint (arrow). There is at least one third subluxation of the head of the metacarpal and only small osteophytes.

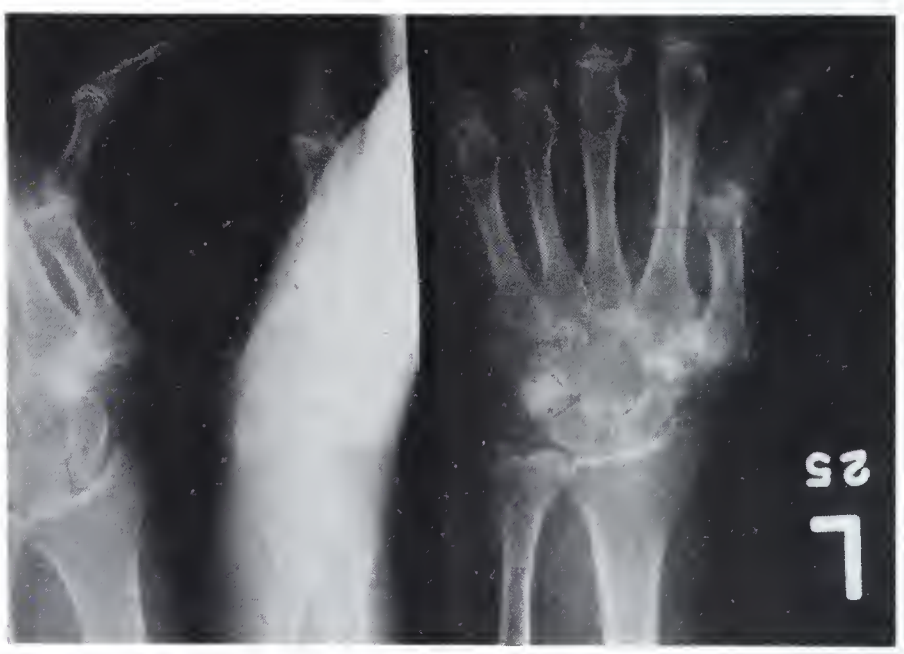

Figure 4: Stage IV radiographic degenerative changes. There are degenerative changes evident at all trapezial facets, with severe subluxation of the thumb metacarpal, large osteophytes, and a very narrowed joint space.

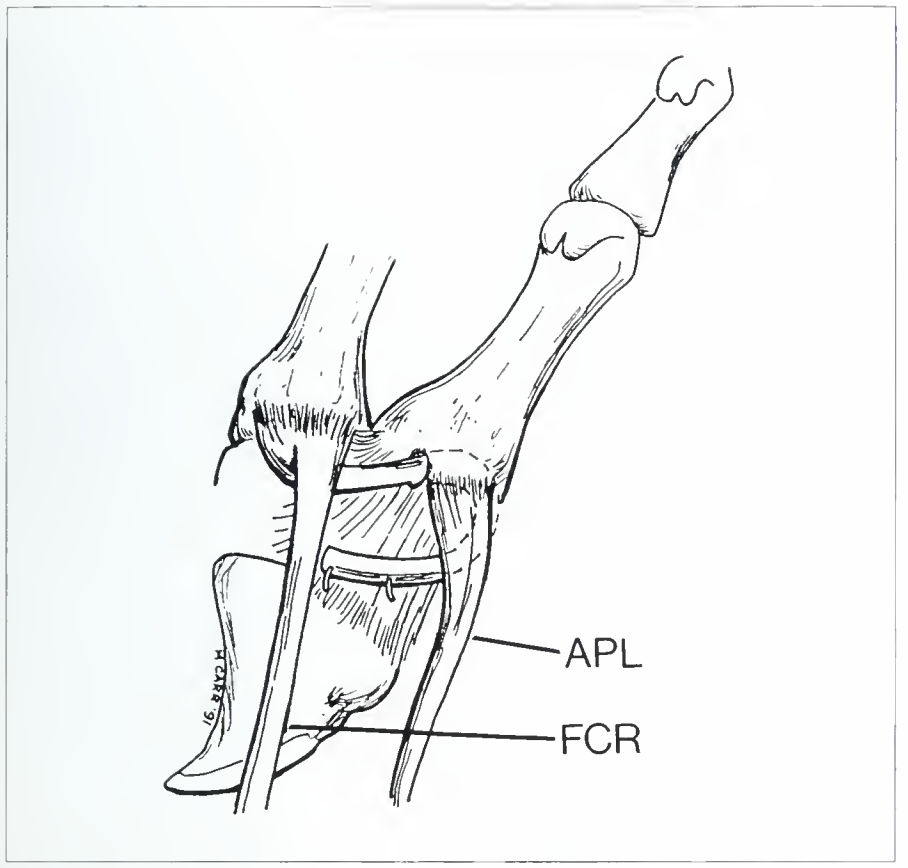

Figure 5: Diagram illustrating the FCR (flexor carpi radialis) ligament reconstruction technique, from the volar aspect. The FCR tendon is split longitudinally leaving the distal end intact. The free end is routed through a hole gouged in the proximal part of the thumb metacarpal, then burrowed under the APL (abductor pollicis longus) and ultimately reattached to the volar aspect of the capsule. Some operators hook the tendon under the remaining FCR before suturing it down to the remaining soft tissue.

Various types of surgical procedures have evolved over the years to deal with thumb basal joint OA that is refractory to conservative treatment. Ligament reconstruction is used to re-approximate normal joint surfaces[5,16,21-26], while arthrodesis (bony fusion)[8,14,16,27], excision of the trapezium[14,20], or resection with synthetic[14,16,17,20,28-31] or autologous[5,14,32] implant placement are used when joint surfaces are inadequate.

Early in the disease, before cartilage damage has occurred, ligament repositioning to stabilize the hypermobile (but passively reducible) thumb CMC joint can prevent further articular damage[5]. Various tendons have been utilized; the extensor pollicis longus[21], extensor carpi radialis longus[22], extensor pollicis brevis[23], abductor pollicis longus(APL)[24], and the flexor carpi radialis (FCR)[5]. Only the FCR procedure (Figure 5) has had long term follow-up and is currently the favoured procedure[14,16,20,25,33]. The joint is approached by a radial incision and the capsule of the thumb CMC joint is opened initially to check the condition of the articular cartilage since ligament reconstruction should only be considered if the cartilage appears normal. The dorsum of the joint is then exposed and a bony tunnel is made dorsovolarly in the proximal end of the thumb metacarpal. With one or two small transverse incisions starting about 8 centimeters proximal to the distal wrist skin creases, the FCR tendon is partially transected and split longitudinally, leaving its distal insertion intact. This tendon strip is then passed dorsally through the tunnel in the metacarpal, then passed beneath the abductor pollicis longus tendon close to its metacarpal insertion and finally attached back to the remaining FCR on the volar aspect. This pathway strengthens the CMC joint capsule on the volar, dorsal and radial sides[5,10,14,25]. Fixation with a Kirshner ("K") wire and cast for four weeks is followed by physiotherapy to regain hand function[14]. In long term follow-up, there is acceptable stability and pain reduction when the procedure is used in the early stages of the disease[10].

There is ongoing debate over the treatment of advanced CMC degeneration. Arthrodesis using a grafted bone strut between the first and second metacarpals[8] or more commonly fusion across the CMC joint requires prolonged casting and is not always successful[14,16,27]. Pain is resolved but mobility is decreased, so the procedure has been abandoned by many operators. Excision of the trapezium also relieves pain leaving better mobility, but stability and strength are compromised and the thumb is shortened which may be cosmetically unacceptable[28]. Total excision of the trapezium has been advocated for pantrapezial arthritis but recent cadaver studies have shown that more than two surfaces are never involved[13]. Therefore the procedure has become contraindicated.

If only the TM joint is affected and the ST joint is in good condition, the treatment of choice is ligament reconstruction combined with tendon interposition arthroplasty (Figure 6)[14,15,32,33]. The head of the metacarpal and the distal articular surface of the trapezium are resected to create a flat surface perpendicular to the functional axis of the thumb. Ligament reconstruction is begun as previously described but the free end of the FCR tendon strip is transected and used to interpose the resected joint surfaces, either folded as a four layer sandwich[15,33] or rolled like an anchovy[32]. Then the remaining FCR is routed as previously described and sutured to the APL tendon. Fixation is similar to that described for ligament reconstruction. Follow-up of several years has shown significant pain reduction in all patients, while range of motion and pinch strength are equal to the unoperated hand[15,33]. Animal models have revealed that the interpositional tendon graft is replaced by mature scar tissue and metaplastic cartilage[34].

The literature describes a wide variety of synthetic implant designs. Braun described an implant combining a distal metal ball-shaped articulating surface with a proximal polyethylene socket, both of which are cemented into bone[36]. The articulation locks mechanically so that capsular reconstruction is unnecessary[36]. The more common implants are made of

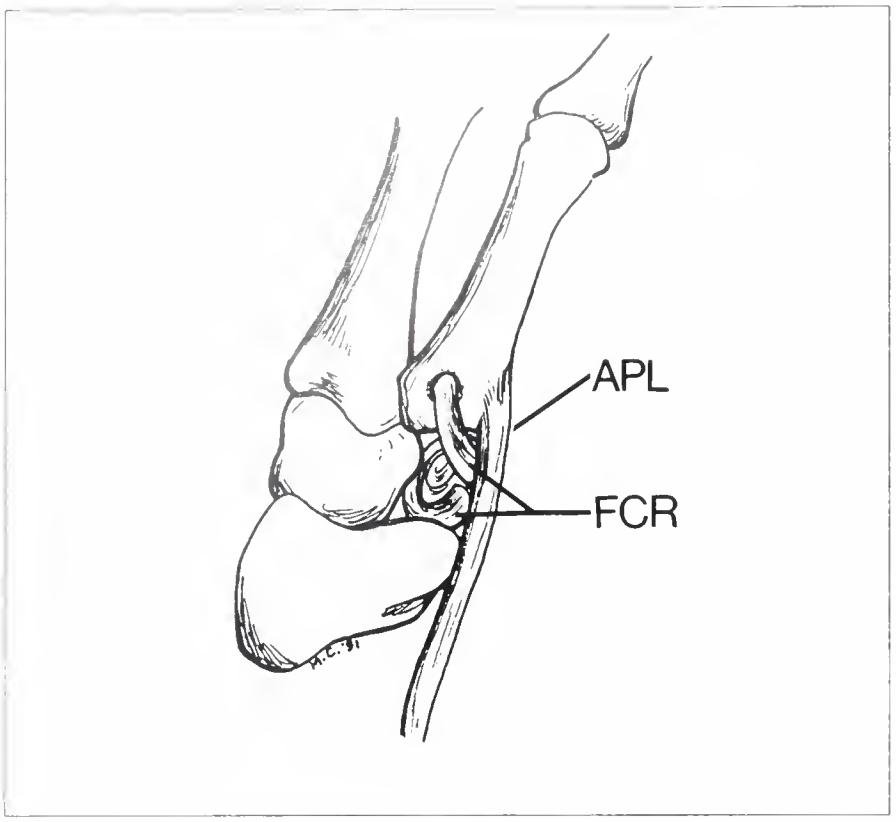

Figure 6: Diagram illustrating the FCR tendon interposition arthroplasty combined with ligament reconstruction, viewed from the dorsoradial aspect. A small segment of freed tendon is first transected and stuffed into the space left by the excised trapezium. The FCR tendon is routed as in Figure 6.

silicone- for example a silicone button has been used as a cushion in the TM joint[16]. Most silicone implants have a stem which is inserted into the metacarpal. The Niebauer implant is unique for the Dacron mesh on its stem which allows tissue ingrowth thus stabilizing the implant[29]. Heads also vary in configuration. A small convex head articulates with a trapezium which is still in good condition[27]. If both TM and ST joints are severely affected the trapezium is replaced with a large headed ("Swanson") implant which articulates with the scaphoid[17,28]. There is a high incidence of subluxation of these implants and therefore various methods of stabilization using tendon reconstruction have been devised to rectify this; one implant has a channel cut through the head for passage of a rerouted APL tendon[31]. Both the Swanson and Niebauer implants rely on the use of the FCR ligament reconstruction to prevent implant dislocation[28,30]. Advantages of implants include good mobility and biomechanical acceptability[16] although a complex and important joint is typically oversimplified[29]. Debilitating pain, weakness and loss of mobility are indications for implants[31]. The technique yields more satisfactory results in the older patient who does not have a high demand for hand function[20], especially since silicone may become brittle, and eventually disintegrate after several years[20]. Stress may induce cold flow and "silicone synovitis", an inflammatory reaction, has been reported[35]. Failed implants are difficult to salvage so patient selection is especially important[20,28].

Summary

Osteoarthritis of the basal joint of the thumb is a common disease which presents along a variable spectrum of pain, reduction of thumb mobility and joint deformity. It remains a disease in which management is based on clinical assessment. Early disease is managed conservatively with rest, anti-inflammatory agents or analgesics and intra-articular steroid injections. Since the course of OA is often benign, it is important to avoid early invasive treatment except in the presence of proven anatomical derangement. Thus, direct inspection of articular surfaces at the time of operation is necessary to decide which of the many available surgical procedures is most appropriate.

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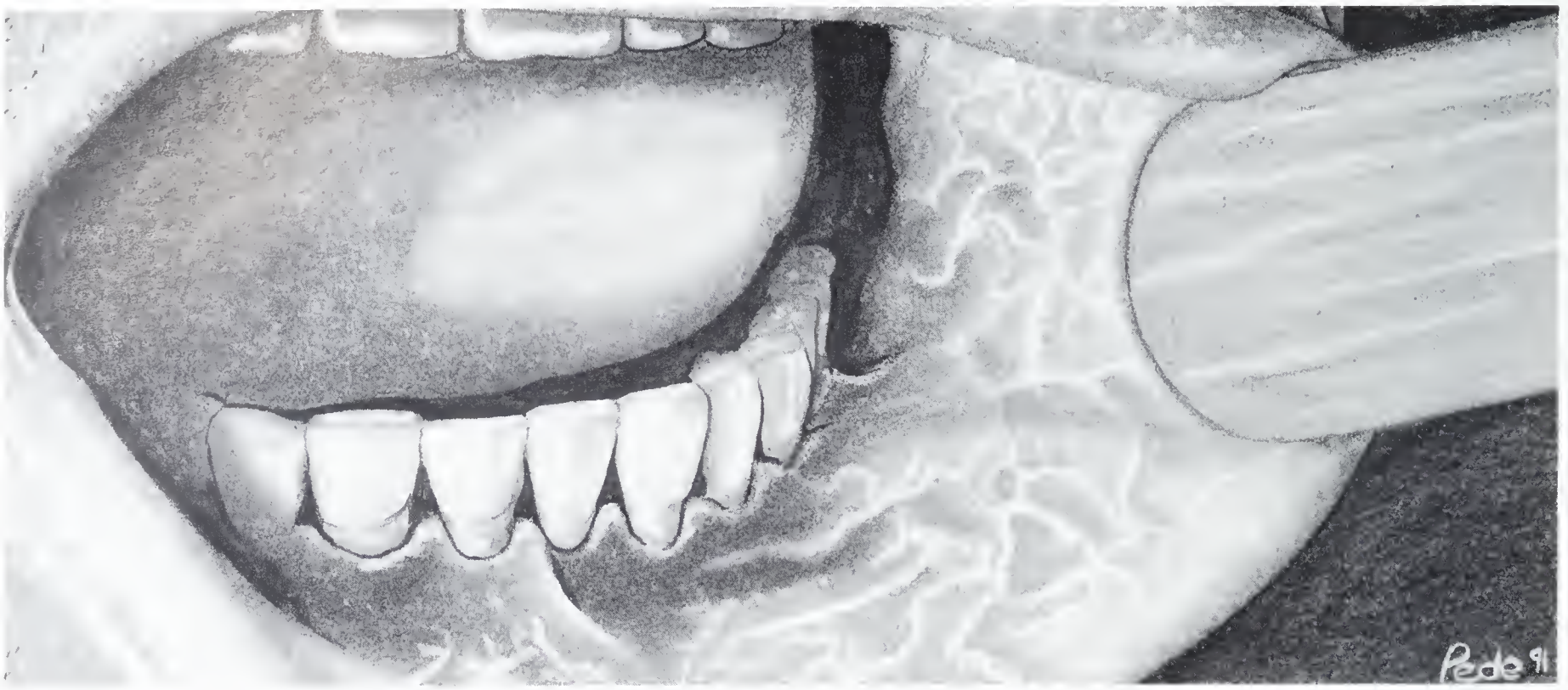

Oral Lichen Planus: Clinical and Pathological Perspectives

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Abstract

Oral Lichen Planus is a disease of the oral mucosa related to the cutaneous form of lichen planus. It has six typical appearances, and its presentation ranges from asymptomatic to extremely painful. Various factors have been proposed to be of etiological significance, including immunologic disease, liver disease, infection, diabetes, hypertension, pyschogenic phenomena, tobacco consumption, and various medications. A malignant potential has been frequently suggested but is still a matter of debate. Treatment involves behavior modification, various medications including topical corticosteroids and a cyclosporine rinse, and surgery in refractory cases.

Introduction

Lichen planus was described as early as 1869, when Dr. Erasmus Wilson reported on the signs and symptoms of a dermatologic condition affecting both the skin and the oral mucosa.[1] The prevalence of lichen planus (LP), in different study populations including dermatology clinic patients, dental patients, and the general population has been cited as ranging from 0.1% to 1.2%[2]. A recent study of 23,616 white Americans over the age of 35 reported the prevalence of oral lichen planus (OLP) to be 1.1/1000[3]. Most patients with cutaneous lichen planus develop OLP some time in the course of their disease, and the oral lesion may precede the cutaneous lesion. Scully cites a study of almost 200 patients with lichen planus which demonstrated 40% with both cutaneous and mucosal involvement, 35% with cutaneous LP alone, and 25% with only mucosal LP[2]. Another study suggests that 30% of patients with oral lesions have signs of cutaneous involvement[5]. Lichen planus is a panracial disease, and is generally felt to have a slight predilection for females. While OLP occurs primarily in individuals over the age of 30 and is rare in children, it can affect any age[5].

Presentation

Cutaneous lichen planus presents itself as intensely pruritic, violaceous flat-topped papules. These papules may be overlaid by a fine-lacy white network of striae known as "Wickham's Striae," which bear a resemblance to the lichen that grows on rocks or stones. The condition may manifest as a single lesion, but multiple to hundreds of lesions are more common. The flexor surfaces of the extremities, particularly wrists and elbows, are common sites, as is the abdomen. The glans penis is a less common site. The cutaneous lesion usually resolves one to two years after onset, but often leaves post-inflammatory hyperpigmentation and atrophy.

Oral lichen planus can be an incidental, asymptomatic finding upon dental examination, or a symptomatic lesion that is painful, hypersensitive, or rough-feeling to the patient. One quarter of patients with OLP have few or no symptoms[7].

Six clinical appearances of OLP have been described, and more than one may be present at one time. They are as follows:

1. Reticular: This is the usual variant, and often accompanies some of the others. It is characterized by finely elevated interlocking "Wickham's Striae," which are less commonly known as "Honiton Lace". The buccal mucosa is affected most frequently (Figure 1).

2. Papular: These are 0.5 - 1.0 mm whitish elevated lesions that appear singly or in groups, and may present in clusters giving rise to a pebbled appearance. They sometimes coalesce (Figure 2).

3. Placque-like: These are white, coarsely textured lesions that are often included in the diagnosis of leukoplakia. They may be surrounded by reticular lesions (Figure 3).

4. Atrophic: These are inflamed areas with reddened, thin-looking mucosa resulting from atrophy of the epithelium. They may be surrounded by reticular lesions or striae (Fig. 4).

5. Erosive (Ulcerative): These lesions are characterized by ulcerated mucosa with a depressed central portion, and are usually surrounded by reticular radiations. They most commonly occur on the tongue and buccal mucosa, and may closely resemble the lesions of pemphigoid, pemphigus, or lupus erythematosus. Gingival involvement may result in desquamative gingivitis (Figure 5).

6. Bullous: Bullous OLP is rare, and is characterized by small bullae or vesicles ranging in size from several millimetres to several centimetres. These lesions may precede erosive OLP (Figure 6).

Erosive, atrophic, and bullous forms are painful. Reticular and atrophic OLP are generally regarded as being the most common clinical presentations, and bullous and erosive as being the least common. Many studies are in agreement in ranking the sites of lesions, from most common to least common, as buccal mucosa, tongue, gingiva, palate, labial mucosa, floor of mouth[2]. Lesions usually occur bilaterally.

Course of Cutaneous and Oral Lichen Planus

The natural history of the oral and cutaneous forms of lichen planus are quite different. While the cutaneous lesions may persist for over a decade, approximately two thirds of patients with this form of lichen planus will experience spontaneous remission after approximately fifteen months[2]. Oral lichen planus generally follows a much longer course. Thorn et al[8] reported papular lesions may vanish within the first year of observation and ulcerative lesions generally persist for only slightly longer, but the plaque-like and atrophic forms are diseases characterized by the persistence and development of additional lesions. In their prospective study of 611 patients, the authors demonstrated complete remission in only 17% of patients, after a mean study time of 7.5 years. Reticular forms may persist for more than twenty years.

Histopathology

The histopathological features of cutaneous lichen planus were described as early as 1909[9]. These features are similar to

Table 1 : Incidence of the most common histopathologic findings in biopsy specimens from 112 patients with Oral Lichen Planus.

	Finding	Incidence
1)	Mononuclear Infiltrate	100%
2)	Lymphocyte Predominance in Infiltrate	99%
3)	Parakeratosis	93%
4)	Basal Layer Degeneration	84%
5)	Prominent Granular Layer	83%
6)	Acanthosis	79%
7)	Intracellular Edema	62%
8)	Widening of the Basement Membrane	61%
9)	Intercellular Edema	57%
10)	Areas of atrophy and Hyperplasia	51%

(Modified From: Hedberg N, Ng A, Hunter N. A Semi-quantitative assessment of the Histopathology of Oral Lichen Planus. J Oral Pathol 1986: 15: 268-272)

those of cutaneous LP, but not as consistent. There is controversy as to the histologic criteria characteristic of, and necessary for, a diagnosis of OLP. Hedberg et al[10] performed a retrospective study of 112 cases in an attempt to establish which parameters are useful in making a histopathological diagnosis of OLP. In an assessment of biopsy specimens from different areas of the oral cavity, and of all clinical forms of OLP, they concluded that lymphocyte infiltration beneath and adjacent to the epithelium, and degeneration of the basal layer were consistent findings. Parakeratinization, acanthosis, and a prominent granular layer were also frequent findings (Table 1).

Immunopathology

An autoimmune basis to lichen planus has long been postulated. The epidermal histological changes in OLP are very similar to those of graft versus host disease. The first observable change is the increased prominence of (antigen-processing) Langerhans cells, even before leukocytes appear in the lamina propria, and when basal changes are not yet notable[11,12]. The identity of a specific antigen responsible has not yet been fully established. Olson et al[13] have demonstrated a Lichen Planus Specific Epidermal Antigen (LPSA) in the granular and deep prickle cells of skin afflicted with cutaneous LP, and the presence of specific antibody to LPSA in LP patients. In "drug-induced" oral lichenoid lesions, the offending substance may result in the specific antigen becoming more apparent to the immune system. As Hedberg et al[10] note, the discovery of HLA-DR (MHC II) alloantigen expression on epithelial cells in lichen planus strengthens the argument for a contact-hypersensitivity type of reaction. (The expression of this "Group D" histocompatibility antigen, which is normally restricted to Langerhans cells of bone marrow origin, means that the epithelium can function in antigen presentation.) The mononuclear infiltrate in the lamina propria is primarily a T-lymphocyte infiltrate, and the fact it contains only a few lymphoblasts, B-lymphocytes, mast cells, or macrophages indicates the reaction is primarily one of cell-mediated immunity.

The pathological reaction has been postulated to proceed as follows[11]: Langerhans cells process the antigen and present it to T-lymphocytes, which in turn destroy keratinocytes. The epidermis then responds to the destruction of the keratinocytes by regeneration both within the lesion and at the periphery. Thus, fully developed lesions of LP have features of both epidermal destruction (lymphocytic infiltrate,

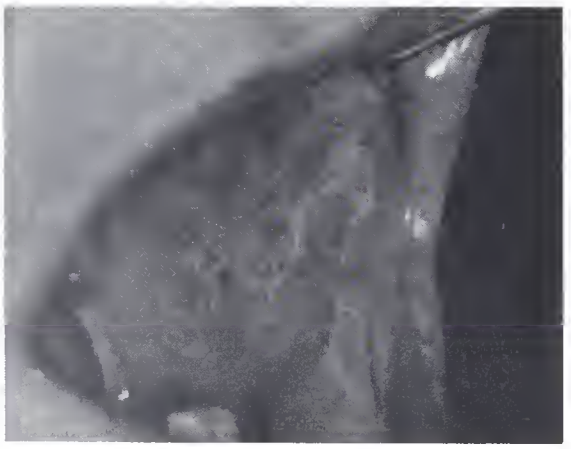

Figure 1: Reticular

Figure 2: Papular

Figure 3: Plaque-like

Figure 4: Atrophic

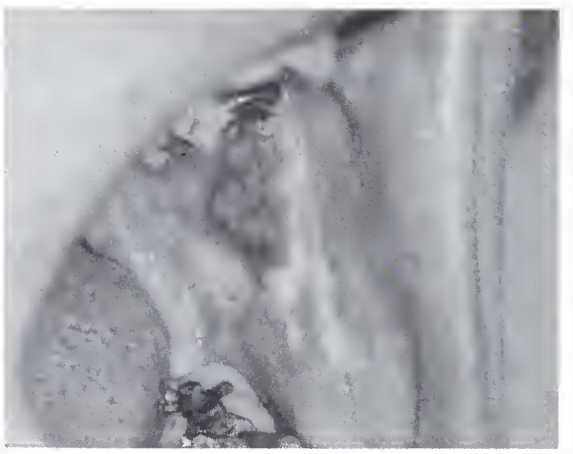

Figure 5: Erosive (Ulcerative)

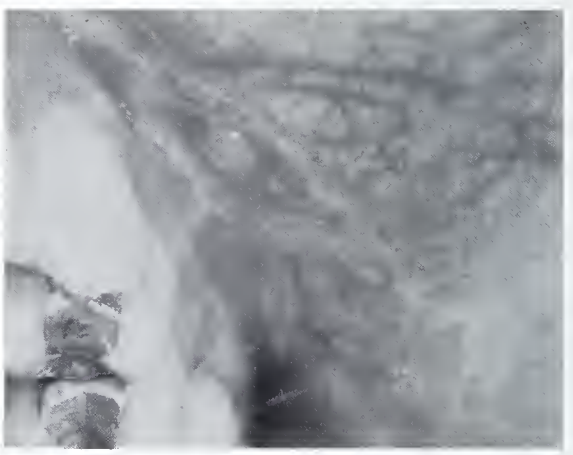

Figure 6: Bullous

vacuolar degeneration along the basal layer, necrotic keratinocytes) and regeneration (mitoses, irregular epidermal hyperplasia, hypergranulosis, and hyperkeratosis.) Direct immunofluorescence reveals fibrin below the basement membrane in nearly all cases, complement in most, and variable IgG and IgM immunoglobulin deposits at the basement membrane zone[2]. These inconsistent results are generally not interpreted as being evidence of a humoral response, but of nonspecific secondary events following damage of the basal layer[10]. Hedberg adds that the T cell nature of the mononuclear infiltrate does not support a humoral response[10].

Etiological Factors

1. Immunologic Disease: As already noted, there is strong evidence supporting an immunologic component to OLP. Lichen planus has been noted to occur with various autoimmune diseases, including lupus erythematosus, pemphigoid, dermatitis herpetiformis, alopecia areata, vitiligo, ulcerative colitis, myasthenia gravis, and Sjogren's syndrome. However, no definite pathologic relationship has been established. These uncommon associations may be coincidental, or resulting from inaccurate diagnosis owing to overlap in diagnostic features (for example, with discoid lupus erythematosus)[14].

2. Liver disease: A number of investigators have noted a correlation between lichen planus and liver disease, including chronic active hepatitis and primary biliary cirrhosis. Reborah and Rongioletti[15] reported that 11.3 of 44 patients (27%) with chronic active hepatitis had evidence of LP. Associations noted have often been unaccompanied by a demonstration of an actual coexistence of both disease processes. Skully describes a study whereby 8% of 113 patients with LP had at least one abnormal liver function test, but no patient exhibited unequivocal biochemical evidence, such as serum autoantibodies, nor clinical stigmata of liver disease.[16] At present, while the association of lichen planus with autoimmune liver disease is probably more than coincidental, this relationship will for the time being have to be categorized as theoretical.

3. Infection: A bacterial etiology to lichen planus was originally hypothesized. Different studies have reported microbial findings including gram negative anaerobic bacilli, rod-like bodies resembling bacteria, and spirochaetes; none of these results have been confirmed[2]. A postulated viral etiology fell out of vogue when electron microscopy (EM) and viral isolation studies failed to demonstrate the existence of a virus[2]. Olson et al[13] have recently theorized that the restriction of LPSA to to skin tissues of LP patients, combined with the finding that approximately 7% of sera from non-lichen planus people contain antibody to LPSA, supports a possible infective etiology of LP.

Table 2 : Distribution of 722 patients with Oral Lichen Planus and the study population according to tobacco habit and sex

	Men		Women		Total	
	Study Pop'n	OLP	Study Pop'n	OLP	Study Pop'n	OLP
No Habit:	1,393	5 0.4 %	5,110	45 0.9 %	6503	50 0.8 %
Smoker:	9,040	89 1.0 %	195	2 1.0 %	9235	91 1.0 %
Tocacco Chewer:	2,270	120 5.3 %	6,222	312 5.0 %	8492	432 5.1 %
Smoker & Chewer:	3,249	144 4.4 %	121	5 4.1 %	3370	149 4.4 %

(Modified From: Murti PR. et Al. Malignant Potential of Oral Lichen Planus: Observations in 722 Patients from India. J Oral Pathol 1986: 15: 71-77.)

4. Diabetes and Hypertension: It has been hypothesized that colloid or "Civatte" bodies develop as a result of damage to the epithelium caused by hypoxia or anoxia subsequent to circulatory disorders[18]. Such disorders could be secondary to hypertension or diabetes mellitus. (Similarly, damaged blood vessels may release a substance promoting hyper-keratinization.) The triad of OLP, hypertension, and diabetes mellitus has been described as Grinspan's Syndrome[19], but further studies by Christenson et al[20,21] have failed to support an association beween lichen planus and either of the latter two conditions.

5. Psychogenic Factors: A psychogenic etiology was suggested as early as Erasmus Wilson's paper, in which many of his 50 patients were described as "highly hysterical" or of "highly nervous temperament"[1]. More recent papers have suggested a loose association between OLP and psychiatric disturbance, including a comparison by Hampf et al[22] of 56 patients with verified OLP to normal controls. The study suggested a statistically greater incidence and severity of "mental disturbance" in patients with OLP. Patients often reported that the clinical appearance of OLP, including discomfort, became worse during times of emotional stress. However, none of the patients reported a subjective feeling of mental disturbance, and only one felt the need for psychiatric treatment. A review of the literature suggests that while emotional factors may promote exacerbation of lesions, there is no evidence for a psychogenic etiology to OLP.

6. Tobacco: The hypothesis that smoking is linked etiologically to OLP is essentially refuted by a study of 611 patients with OLP which concluded that patients with OLP smoke cigarettes no more than controls[23]. It is suggested by this report and others that plaque-type afflictions are more frequent among smokers than among nonsmokers. Consumption of tobacco, though, may still be linked to OLP. A study in India surveyed 27,599 individuals, 722 of whom had OLP. While there was no marked difference between the prevalence of patients with OLP among non-smokers and smokers, those who chewed tobacco were at five times increased risk over the general population (Table 2)[24]. As with many past studies linking OLP and tobacco, however, this large and recent study may also be flawed. The fact that only 90% of patients biopsied demonstrated a band-like infiltrate suggests other conditions, such as leukoplakia, may have been mistaken for OLP.

7. Medication: Lichenoid eruptions of the mouth and skin have been reported in association with a multitude of drugs. It has been theorized that the fact there are well documented cases of chemically unrelated drugs causing recurrence of OLP lends support to the notion that lichenoid drug reactions are not allergic reactions, but that drugs may unmask or promote OLP[14]. Moreover, it is felt that some of the disease associations with OLP may in fact be attributable to the medications used to treat them, including oral hypoglycemic agents for diabetes and d-penicillamine for autoimmune diseases[25].

Well over 25 drugs have been implicated with lichenoid lesions (see table 3). A study by Potts et al[26] comparing medications taken by 225 patients with OLP to 202 age and gender-matched controls treated at the same clinics demonstrated no significant differences in the use of various classes of medications, such as oral hypoglycemic agents and psychotropic drugs, between the two populations. However, 17% of the patients with lichen planus were taking NSAIDs, compared with 9% of the controls, and 30% of patients with erosive OLP were taking NSAIDs, compared with the 9% of patients on NSAIDs who had the other clinical variants. The relationship between NSAIDs and erosions was proven by drug withdrawal and challenge tests. Moreover, histological examination distinguished these lesions from nonspecific mucosal ulceration also associated with these drugs. In at least one third of the OLP patients, the history was strongly suggestive of drug ingestion followed by the onset of OLP. Overall, the results strongly suggest OLP may be aggravated or precipitated by nonsteroidal anti-inflammatory drugs, and that withdrawal of the drug may benefit some patients[26]. (This study, however, also produced some inconsistent findings; the authors note that fewer OLP patients were taking antihypertensive medications than controls; $p < .05$.)

The Malignant Potential of Oral Lichen Planus

The question of whether OLP has definite premalignant potential has been a longstanding controversy that has not yet been satisfactorally resolved. The incidence of neoplastic transformation of OLP, in different studies, ranges between 0.3% and 3.0%; however, figures between O% and 10% have been cited[27]. There are myriad factors which can confound the problem and lead to an erroneous conclusion of neoplastic transformation. Some case reports did not clearly indicate a correspondance between the site of the malignant lesion and the original OLP lesion. Others have used histologic criteria that are not adequately rigid to distinguish OLP from premalignant epithelial dysplasia. Moreover, the possible coincidental occurrence of both of these relatively common conditions cannot be discounted. Krutchkoff et al[28] reviewed 222 malignant transformations cited in the literature, and concluded that only 7% were acceptable by their criteria. The remaining cases were excluded on the basis of 1) insufficient evidence to support the diagnosis of lichen planus, 2) the occurrence of cancer in a site remote from the lichen planus, or 3) inadequate historical data regarding past exposure to carcinogens. Eight of the fifteen remaining patients with oral cancer had associated known carcinogenic risk factors. It was concluded that the evidence was insufficient to demonstrate that OLP poses an inherent biologic potential to turn into cancer, but that the risk of developing oral carcinoma is slightly increased in patients with OLP.

Murti et al[24] assessed the malignant potential of OLP on the basis of observations of 722 patients with OLP. These subjects were detected in three population-based, house-to-house studies examining a total of 27,599 individuals in India. The 722 patients with OLP were re-examined annually over a ten-year period with a mean observation period of 5.1 years. OLP was diagnosed on a clinical basis when the lesion showed Wickham's Striae. Biopsies were taken whenever patients consented (13%). Carcinoma was noted to develop in three (0.4%) patients with OLP, all of whom were smokers. The relative risk of a smoker with OLP developing oral cancer, compared to that of a similar smoker without OLP, was calculated to be 3.3. This relative risk, however, failed to reach significance level, assuming Poisson distribution for the occurrence of the three oral cancer cases. Moreover, a careful reading of the original article suggests that only one of the three patients had histological proof of prior OLP. The authors concluded their findings could not substantiate with any high degree of certainty a significant potential for the malignant transformation of OLP[24].

Table 3 : Drugs having been implicated with lichenoid lesions

Amiphenazole
Arsenic / Bismuth
Captopril
Carbamazepine
Chloroquine
Chlorpropamide
Dapsone
Demeclocycline
Dental Amalgam
Furosemide
Gold
Labetalol
Levamisole
Mepacrine
Methyldopa
NSAIDs
Oxprenolol
Para-Amino Salicylic Acid
Penicillamine
Penicillin (in secondary syphillis)
Phenothiazines
Practolol
Propranolol
Quinacrine
Spironolactone
Streptomycin
Tetracycline
Thiazides
Tolbutamide

(Modified from: Skully C., El-Kom M. Lichen Planus: Review and Update on Pathogenesis. Journal of Oral Pathology 1985: 14, 431-458)

A more recent Danish study following 611 patients with OLP for a mean of 7.5 years each produced more conclusive results. During follow-up, nine patients (1.5%) developed oral squamous cell carcinoma (SCC) in areas of previously diagnosed OLP. The incidence of smokers with carcinoma among the OLP group was felt not to differ from that of the general Danish population. The estimated number expected to develop oral cancer in a sample of the general Danish population that was of similar size, age distribution, and follow-up was 0.18. The considerable increase of oral carcinoma among OLP patients compared with controls was statistically significant ($p < 0.00001$). This study, though, was also not without potential flaws. For example, it was based on patients who were referred to a hospital oral medicine and oral surgery clinic, thus forming a highly selected group[29].

Essentially, a review of the literature leads to the conclusion that there is insufficient evidence to demonstrate that OLP is a clearly premalignant condition. However, various subgroups of patients with OLP - namely those with atrophic or ulcerative lesions, as well as chewers of tobacco warrant careful and regular follow-up.

Treatment

The presence of OLP is not in itself an indication for intervention; only if irritating to the patient or ulcerative should this condition be treated. The aim of treatment is twofold. Firstly, it should provide symptomatic relief of a potentially distressing condition. Moreover, it should diminish the likelihood of this potentially precancerous condition becoming malignant. Few studies have been performed to assess whether the latter aim is achieved by treatment.

An early step in treatment is to smooth off rough teeth. As some postulate that lichen planus is a disease affected by the Koebner phenomenon, the so-called "isomorphic response" where injury to uninvolved skin results in acceleration of epidermal growth, reducing trauma may help reduce the oral form as it helps the dermatologic skin form[14]. Thus, it is important to discourage trauma-inducing habits such as cheek-sucking and tongue-rubbing, and oral habit splints may be necessary to control these tendencies.

A careful drug history must be taken from these patients. As already noted, a plethora of drugs have been hypothesized to trigger or unmask OLP, and thus no drug can be beyond suspicion. Within the limits of safety, the physician should assess whether withdrawal of suspicious drugs results in at least some resolution of the lesion.

The mainstay of treatment in OLP is topical corticosteroid therapy. Various techniques have been employed to keep the medication applied to the lesion, including having the patient hold a medication-laden gauze pad against the lesions, as was first suggested by Birt et al[30]. Application of the ointment under plastic wrap wound around the tongue has been used for glossal lesions. Treatment is usually administered for 20 to 30 minutes on a TID or QID basis. Commonly used preparations include betamethasone valerate 0.1% and triamcinolone acetonide. The possibility of adrenal suppression secondary to longterm steroid use cannot be ruled out.

Intralesional injections of corticosteroids, including hydrocortisone and dexamethasone sodium phosphate, have been used to treat erosive lesions unresponsive to topical therapy.

Other treatments have been tried and found to be less desirable, owing to their being less effective and sometimes potentially toxic. These have included topical isotretinoin[31],

systemic isotretinoin[32], and griseofulvin[33]. Limited success has been reported with oral psoralens and ultraviolet A (PUVA) directed toward the oral lesions[34].

A recent report by Eisen et al[35] showed marked improvement on topical cyclosporine. After eight weeks of using a cyclosporine mouthwash (swished and expectorated) on a TID basis, the eight test patients showed marked improvement in erythema (P=0.003), erosion (P=0.02), reticulation (P=0.007), and pain (P=0.002), and commonly experienced remissions lasting from three to eight months. The eight control patients (having received the cyclosporine vehicle only) had no change or minimal improvement, but improved in a fashion similar to the test group when administered the cyclosporine treatments. The authors add that there is evidence topically applied cyclosporine may be an economical alternative to the mouthwash rinses, which cost significantly more than topical steroids.

Cryosurgery has been reported as providing longstanding relief for lesions refractory to medical therapy[36].

Lastly, while the extent to which OLP is a premalignant condition is still subject to debate, it is clear dysplastic areas can be mistaken for lichenoid regions, and also that OLP and oral cancer can occur simultaneously. For this reason, patients with OLP should be followed at three- to six-month intervals, and any suspicious lesion should be biopsied. Biopsy is also indicated for lesions refractory to medical treatment, lichenoid lesions in atypical sites for OLP (eg. floor of mouth), and lesions in patients with a history of oral carcinoma.

Conclusion

Oral lichen planus is encountered frequently by the general practitioner and is seen fairly often by dermatologists, dentists and otolaryngologists, particularly those participating in mouth clinics. Moreover, the importance of this condition stems not only from its clinical significance, but also from the fact it is of academic interest, as an evolving condition of unknown and possibly multifactorial etiology. The intrigue surrounding this disease brings to light the unfortunate truth - that possibly the majority of studies looking into the etiology of OLP or its premalignant potential are imperfect investigations. It is the impression of the reviewer that further rigorous studies are needed to make concrete conclusions regarding this fascinating condition.

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A Case of Acute Abdominal Pain

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Mr. C., a 22-year-old white male from Toronto was brought to the Emergency Room by his wife on August 20, 1990 complaining of an excruciating pain in the right lower quadrant of his abdomen which woke him up that morning. He had always been in good health up until that morning. The pain had a sudden onset, was colicky in nature and was aggrevated by any type of movement, and did not radiate to the back. He felt nauseated and vomited before he left home. Remarkable on physical examination were: Temp. 38.5 °C; heart rate: 96/min regular; respiratory rate 25/min. Abdominal: right lower quadrant (RLQ) abdominal tenderness on coughing; guarding and rebound tenderness referred to RLQ; positive McBurney's sign; palpable mass in RLQ.

Question: What is your diagnosis and how should Mr C. be treated?

Answer: All the above symptoms and signs are classical for an acute appendicitis. Appendectomy is the only acceptable treatment for suspected acute appendicitis. The differential diagnosis should, however, include the following disorders: viral gastroenteritis; salmonellosis; Yersinia ileitis; pseudomembranous enterocolitis; Crohn's disease and right ureteral calculus.

The appendix was found to be normal during laparotomy. The terminal ileum was inflamed. The cecum and the base of the appendix were not involved in the inflammation. Given the site of the inflammation, a diagnosis of acute (uncomplicated) Crohn's disease was made.

Question: What would you do next? Resect the affected part of the ileum and/or the appendix or neither?

Answer: Resection of the ileum is not indicated, since acute (uncomplicated) Crohn's enteritis may become asymptomatic for an extended period of time without recurring or progressing to chronic Crohn's Disease. The appendix should be removed in order to prevent the problem of differentiating between Crohn's disease and acute appendicitis following a recurrent attack of cramping abdominal pain.

Mr. C was discharged from the hospital after the appendectomy. He returned to the Emergency Room one month later with a persistant and excruciating RLQ abdominal pain which started a week after he was discharged. He felt nauseated and had bloody diarrhea for over a week. Weight loss was approximately eight kilograms.

Question: What diagnostic test would you perform to estimate the extent of his intestinal inflammation?

Answer: The best tests are antegrade and retrograde barium studies of the small bowel and large bowel respectively. Also, colonoscopy is required generally to assess colonic involvement.

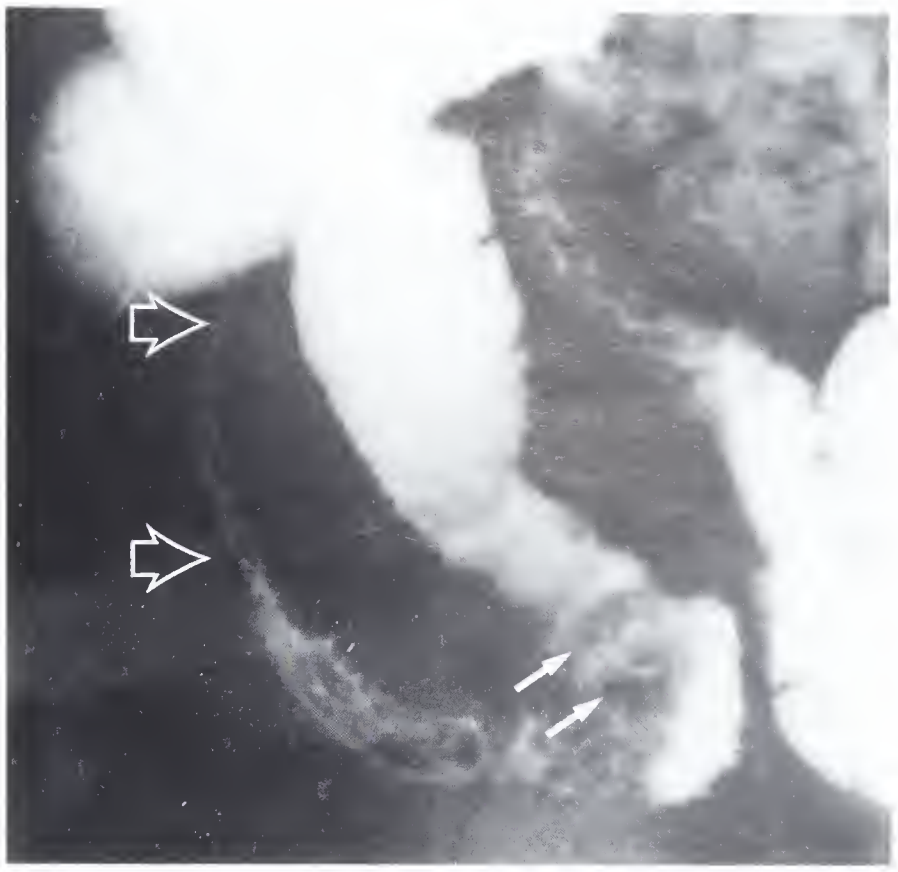

Figure 1 shows the terminal ileum after a barium follow-through study.

Question: What features on this X-ray are suggestive of Crohn's disease?

Answer: Closed arrows: terminal ileum with edematous (due to inflammation) mucosal folds. The last several centimetres (open arrows) are narrowed by spasm and edema (string sign). The loops are seperated because of mural and mesenteric thickening. Other characteristic features of Crohn's ileitis on X-ray (not seen in this case) are: aphthous ulcers, linear ulcers, deep ulcers or sinus tracts, "cobblestoning", fistulae, stricturing and dilatation.

Resection of diseased bowel is indicated in stricturing complications of ulceration such as sinuses, fistulae, abscess formation, perforation into the abdominal cavity, hemorrhage or in cases of nonresponsiveness to medical treatment. Because of the debilitating effects of the ileitis on Mr C., it was decided to resect the affected parts of his ileum and cecum.

Question: What is the prognosis for Mr. C?

Answer: There is no cure for Crohn's disease. Recurrence of the disease is very likely. Medical management (with corticosteroids, sulfasalazine, immunosuppressive drugs such as Imuran, nutritional therapy) and surgical management (resection and bypass) may suppress exacerbations of the inflammatory process. Recurrance almost always occurs at the neo-terminal ileum and reoperation is required in approximately 40 per cent of all patients after the first resection.

Treatment of Isolated Systolic Hypertension

Isolated Systolic Hypertension (ISH) has been defined as a systolic pressure greater than 160 mm Hg and a diastolic pressure less than 90 mm Hg. Its prevalence has been estimated at six per cent in individuals aged 60 to 69 years, and up to 18% in those aged 80 or older. The Framingham Study identified it as the single greatest risk factor other than age for the development of cardiovascular disease in the elderly.

In a recent multicentre, randomized, double-blinded, placebo-controlled trial involving 4,736 male and female patients, researchers found significant decreases in total strokes in the group treated with antihypertensives. The results of the five-year trial, called SHEP (Systolic Hypertension in the Elderly Program), were published recently in a JAMA article "Prevention of stroke by antihypertensive drug treatment in older persons with isolated systolic hypertension". Patients were treated with step-care therapy involving chlorthalidone (a sulfonamide diuretic) or atenolol (a cardioselective beta-blocker) in order to maintain a blood pressure below 160 mmHg for the duration of the trial. Statistical analysis of the data revealed a 36% decrease in stroke incidence (p=.0003), 27% lower incidence of nonfatal myocardial infarction and coronary death, as well as a statistically insignificant 13% decrease in all-cause mortality. Side effects included increased incidence of hypokalemia, hyperuricemia, hyperglycemia and hypercholesterolemia.

SHEP is the first trial to study treatment of ISH. Previous studies, including the Hypertension Detection and Follow-up Program trial, have shown a 42 per cent overall reduction in stroke incidence with treatment of diastolic hypertension. This is similar to that found in the SHEP trial, suggesting the broad effectiveness of antihypertensive drug therapy.

Smokeless tobacco use and tobacco ad bans

Recently a Quebec Superior Court judge reversed the Tobacco Products Control Act on constitutional grounds. This action stirred the old debate on the effectiveness of tobacco ad bans on reducing consumption. Mark Taylor, president of the Addiction Research Foundation, believes this is a step backwards. He stated that "At least 27 other countries have adopted this kind of legislation. We should not be the first to take a backwards step." Taylor also argued that whether or not the issue is good law, it is bad public policy.

Meanwhile, reports in the U.S. show disturbing increases in use of smokeless tobacco, especially among young males. Dr. Louis Sullivan, U.S. Secretary of Health and Human Services, is insisting smokeless tobacco be viewed on par with cigarette smoking as a serious health hazard. Thirty thousand new cases of oral cancer were reported in the U.S. last year, and the numbers seem to be growing rapidly. Twenty six per cent of males between 12 and 17 have tried some form of smokeless tobacco. Statistics in Canada, however, are showing a slow decline in smokeless tobacco product sales. The only exception, sadly, is in young northern Inuit males, whose consumption closely matches that seen in the U.S. Various workers in the field are warning that since Canada still does not have a ban on smokeless tobacco products, concerned Canadians must take appropriate steps soon.

Analgesics and Osteoarthritis

Osteoarthritis (OA) has been hailed as the most common joint disease. Some sources estimate that up to 80% of the population has radiographic evidence of OA by age 65, with approximately 60% of these individuals having symptoms. Standard treatments aimed at relieving joint pain and improving mobility involve the use of NSAIDs, often in high daily doses, even though the role of inflammation in the pathophysiology of cartilage breakdown and consequent joint pain is unclear. In light of the many side effects of NSAIDs, including electrolyte disturbances and gastric ulcer, this practice has been questioned.

In a recent study published in the NEJM titled "Comparison of an anti-inflammatory dose of ibuprofen, an analgesic dose of ibuprofen, and acetaminophen in the treatment of patients with osteoarthritis of the knee", investigators studied the short-term, symptomatic treatment of OA. They compared a 1200 mg (analgesic) dose of ibuprofen, a 2400 mg (anti-inflammatory) dose of ibuprofen, and a 4000 mg dose of acetaminophen in a randomized, double-blinded trial involving 184 patients. The patients were assessed using several scales. The results indicated no significant difference in mean improvements on a pain scale in the three groups. Side effects were minor and similar in all three groups. This study points out the need for more careful, longterm evaluation of the risks and benefits of using various analgesics in the treatment of OA and/or other musculo-skeletal/rheumatological disorders.

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Jablonsky's Dictionary of Syndromes and Eponymic Diseases

2nd edition. Stanley Jablonsky. 680 pages. Robert E. Kriegler Publishing Co. Inc.1991. $99.50

Physicians, like members of most other disciplines, have over the centuries developed a way of communicating amongst themselves in terms which are easily understood by colleagues who are familiar with the subject at hand. The evolution of this "jargon", which is at times referred to in medicine as "medicalese", is seen in many different professions and trades and is a response to two basic needs.

First and foremost is a desire, for the sake of expediency, to avoid unnecessary and long-winded explanations of concepts which are assumed to be well-known by both parties. Secondly, and perhaps less compellingly, there is a tendency to use language which is less comprehensible to the layman, not so much for the purpose of deception or secrecy, but rather to enhance an aura of professionalism.

A perusal of the records which doctors exchange among themselves discloses two favored devices of medical lingo: the acronym and the eponym. Both are ubiquitous in medical literature, often to the frustration of any reader who does not have an encyclopedic knowledge.

Jablonski's Dictionary of Syndromes and Eponymic Diseases is a welcome response to this frustration. Overwhelming in it comprehensiveness, this 700 page tome contains thousands of entries ranging from the painfully obscure to the incredibly mundane.

The dictionary includes entries on both syndromes and specific eponymic diseases. The actual distinction between these two phrases has become somewhat blurred in recent years. The reasons for this, and the significance of the term "syndrome", is explored by the author in his "Historical Note" at the beginning of the text. There he also notes some of the arguments which have arisen within the medical community over the use of personal names in syndrome designations.

The syndromes themselves are listed in alphabetical order and cross-referenced so extensively that one is bound to eventually find the desired entry regardless of the name under which one searches. This work also contains short biographical notes on the men and women whose names have been immortalized through these diseases as well as plenty of photographic illustrations which appear throughout the compendium and thus complement the written text.

Obviously, the nature of this reference text is such that it can never be truly complete since new syndromes are continually being described. Still, despite this inherent difficulty, Jablonski has succeeded in compiling an invaluable, up-to-date reference tool which can be useful for specialists and general practitioners alike.

Deena Ages MD, 9T0

Trials of an Expert Witness: Tales of Clinical Neurology and the Law

Harold L. Klawans MD. 277 pages. Little, Brown & Company Limited.1991. $24.95

It has been my experience that the majority of doctors and medical students view lawyers and indeed the entire legal system with feelings bordering on revulsion or fear. This may seem quite natural given the adversarial positions in which these two professions often find themselves. Still, there are those few brave souls who use their medical knowledge to act as expert witnesses and Harold Klawans, a professor of neurology and pharmacology at Rush University in Chicago, is such a person. Indeed, his involvement has spanned almost two decades and he has now written about some of his more memorable experiences in a new book.

Each chapter in this book looks a one particular neurological problem by recounting the case history of a particular legal action. Some of these stories are fascinating - like the case over whether Jack Ruby, the murderer of Lee Harvey Oswald, had suffered from epilepsy. If this could be proven, then there might be a legal defense for the man who shot the assassin of

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President John F. Kennedy while millions of Americans watched on their television screens. Another action sought to determine whether the trauma of a subway collision could trigger multiple sclerosis in a person. This question cost the New York Transit Authority millions of dollars at the time.

The majority of the stories, however, involve the errors of other physicians. For example, there is the woman who was given D5W (essentially sugar water) intravenously to treat her continuous vomiting due to morning sickness. This order was correct but it was never discontinued and the woman eventually received nine liters in total but was not given any sodium. She eventually went into a coma due to the profound hyponatremia.

Another case involved a women who was misdiagnosed as having Horner's syndrome when she presented with an unopenable eyelid and a dilated right pupil. (A mistake that Dr. Klawan states no competent first-year medical student would make. So, how about it, you first year medical students?) This then led to the overlooking of an aneurysm in the Circle of Willis which subsequently ruptured, causing severe aphasia as well as a weakness on her left side. And these are just the beginning.

Because this new work of non-fiction is targetted for the general public, the text is written in a very non-technical manner and often appears simply to be the trancripts of several conversations. While this can seem a bit odd at times after reading medical texts, it does serve as a change of pace. It also serves as a relatively painless introduction to both basic clinical neurology and medical jurisprudence.

One of the most interesting points which Dr. Klawans brings up is the difference between criminal and civil suits. We are all probably familiar with the famous "beyond reasonable doubt" standard which is required for a criminal conviction. In American civil law, however, there is the "more probable than not" rule. What this means, as one of the lawyers in the book points out, is that it is enough to prove that something is at least 51% probable in order to have it accepted in court. Thus, for those who hope that a plaintiff cannot "prove" anything, this discovery may come as an unpleasant surprise.

Still all this talk about civil suits with huge awards against other doctors raised mixed emotions in me. Some of the mistakes that are documented are so blatant that it almost makes you wince and Dr. Klawans certainly makes you sympathetic to the plight of those who have been harmed by "bad medicine". Still, one begins to wonder if all of us will have to face similar actions because of our honest mistakes. Dr. Klawans is very honest about his opinions:

There is a constant refrain in medical circles that the awards that juries give to patients are far too high . . . There is another refrain equally loud and just as persistent. 'Don't testify for the plaintiffs' . . They are almost always paired with complaints about the high cost of malpractice insurance and the need to practice defensive medicine . . . I always listen politely. Usually that's all I do. Arguing, I have discovered, does not help. It is like arguing religion. Belief in the right of a patient to have legal recourse for an injury resulting from malpractice is like belief in God. It is not a belief that can be inculcated by logical argument. Every time I am tempted to concede to my colleagues may have some degree of justification, I pull out a copy of Mrs. Cook's hospital bills. The award was not sufficient.

Whether one agrees with Dr. Klawans or not, his new book is still an interesting and revealing look into the medical-legal process and is certainly worth your consideration. With the trend toward civil action increasing in Canada, this type of exposure may be very useful in the future and will, at the very least, hopefully spark some interest in these types of issues.

John Lee, 9T3

Clinical Dermatology

3rd edition. Rona MacKee. 364 pages, Oxford Medical Publications. 1990. 29.95 (paperback) $75.00 (hardcover)

The paperback edition of this text is an excellent and affordable introduction to dermatology. Its contents include practical information such as a list of relevant questions for patients presenting with skin lesions, up-to-date descriptions of laboratory tests and plenty of color photographs illustrating each disorder.

The text itself is easy to read with a chapter devoted to each major category of skin disease grouped according to etiology. Each condition is described as to its incidence, etiology, pathology, clinical features, diagnosis and treatment. Points deserving special emphasis are boxed-off from the text and concise tables throughout the book summarize important clinical information.

There is a glossary of useful clinical and histologic terms for those who wish to explore fully the difference between a macule and purpura. And for those who wish to be "on the cutting edge", there are "growth points" which are in essence news flashes detailing recent advances.

All in all, this textbook is well-organized and filled with excellent photographs which may make the purchase of a dermatology atlas unnecessary. Indeed, Clinical Dermatology is worth some consideration from anyone wishing to find a solid introduction to this topic.

Corinne Fischer 9T3

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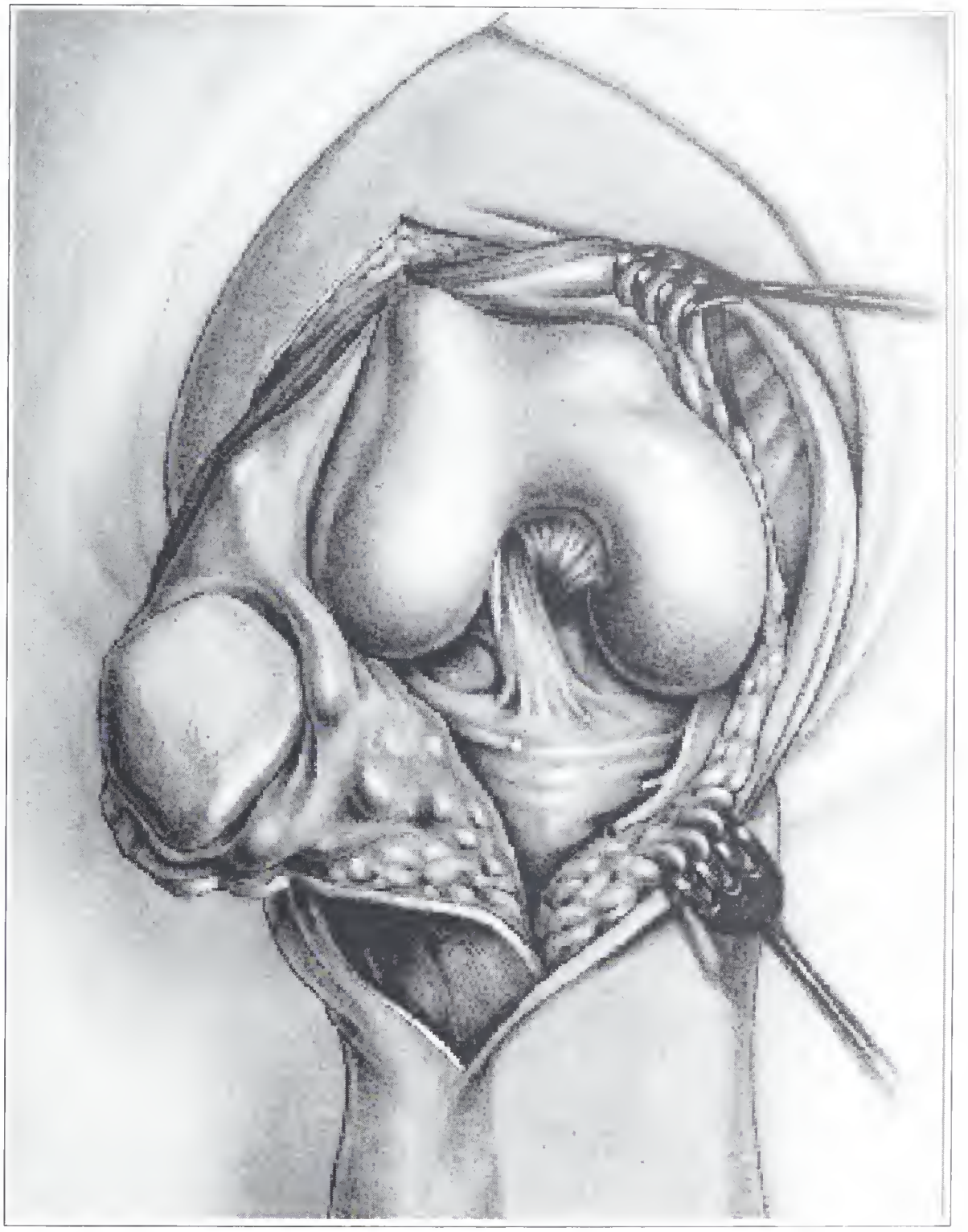

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